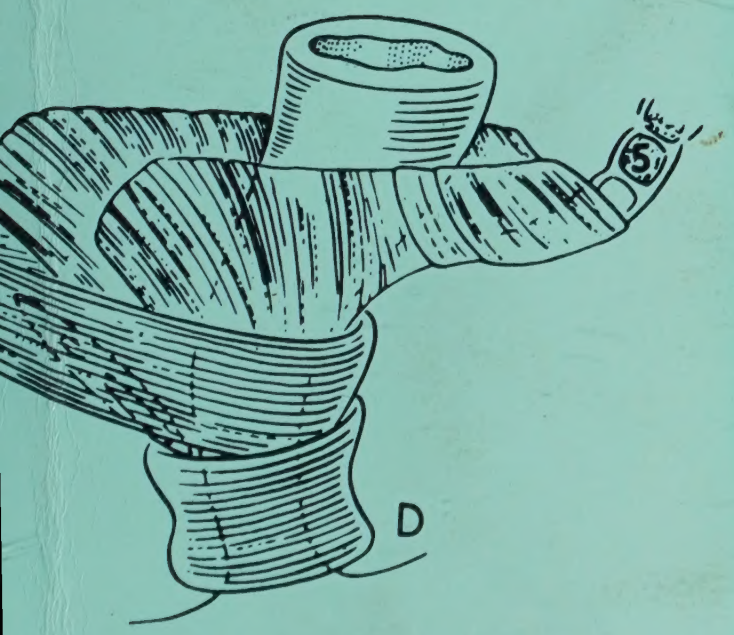
COURSE BOOK



ANORECTAL ANOMALIES
WORKSHOP-1991

Department of Paediatric Surgery
Sri Ramachandra Medical College
and Research Institute, Madras-600 116.

Indian Association of
Paediatric Surgeons

OCTOBER 28, 29, 30, 1991.

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Anorectal Anomalies Workshop - 1991

COURSE BOOK

DEPARTMENT OF PAEDIATRIC SURGERY
THE HOSPITAL FOR SICK CHILDREN
TORONTO

CANADIAN ASSOCIATION OF PAEDIATRIC SURGEONS

October 28, 29, 30, 1991

Anorectal Anomalies Workshop - 1991

COURSE BOOK

DEPARTMENT OF PAEDIATRIC SURGERY
SRI RAMACHANDRA MEDICAL COLLEGE AND
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PORUR, MADRAS-600 116.

INDIAN ASSOCIATION OF PAEDIATRIC SURGEONS

October 28, 29, 30, 1991

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PREFACE

In India each year between 4500 - 5000 babies are born with anorectal malformations (24 million new borns; ARM incidence 1 in 5000 births). The majority of deliveries in India are in the rural areas. These two facts, namely a large numerical incidence and distance from urban specialist care indicate the long way we have to travel to ensure that these malformed babies have the benefit of modern knowledge in the management of Anorectal malformations.

This workshop has been synchronised to be held during the visit of Prof. F. D. Stephens to Madras from October 27 to 30, 1991.

During the second half of the twentieth century, no single person has had such a great influence on surgeons in the understanding and treatment of these malformations than Prof. Stephens. By his papers, books, working groups (Melbourne 1970) (Racine 1984) and innumerable visits to various countries, during four decades, he has moulded surgeons, thinking on the structural abnormality, functional restoration and surgical techniques of correction of these complex malformations. In this endeavour, he has been assisted by able members of the Department of Surgical Research of the Royal Children's Hospital in Melbourne. Notable amongst them have been Mr. E. D SMITH (Co-author of the famous Monograph and its successor Update Book). Mr. ROBERT FOWLER, MISS RUTH MAGNUS and Mr. JUSTIN KELLY.

The Workshop is jointly conducted by the Indian Association of Paediatric Surgeons and the Department of Paediatric Surgery of the Sri Ramachandra Medical College. I would like to thank the Director of the Institute, Prof. N. Madanagopalan, the Dean Prof. C.N. Deivanayagam and the Head of the Dept. of Paediatric Surgery Prof. D. Meera Bai for their generous offer to host the workshop, and Dr. R. Shanbhogue, Associate Professor of Paediatric Surgery and Dr. Kalidasan, Assistant Professor of Paediatric Surgery, for shouldering the principal burden in the organisational aspects of this Workshop.

Grateful thanks are offered to Faculty Members of this workshop and for the registrants some of whom have travelled quite a long distance to participate in this endeavour.

This Workshop is offered as a service to babies afflicted with this malformation in India as well as a tribute to Prof. Stephens and his work.

"WE ARE LIKE DWARFS SEATED ON THE SHOULDERS OF GIANTS. IF WE SEE MORE AND FURTHER THAN THEY, IT IS NOT DUE TO OUR OWN CLEAR EYES OR TALL BODIES, BUT BECAUSE WE ARE RAISED ON HIGH AND UPBORNE BY THEIR GIGANTIC BIGNESS".

BERNARD OF CHARTRES 1119.

Dr. T. Dorairajan

Prof. F. DOUGLAS STEPHENS

Prof. F. Douglas Stephens of Melbourne is one of the world's leading Research Paediatric Surgeons in the fields of Paediatric Urology and Congenital Malformations particularly Anorectal Malformations and their corrections. Now in his 78th year, he is still active in his academic pursuits and is much in demand throughout the world as a Guest Lecturer and Visiting Professor.

He is the third generation doctor in the family and including the career of his father and grandfather the family has been practising paediatric surgery for over a hundred years.

He has trained several foreign surgeons in paediatric surgery and has introduced them to the methodology of paediatric surgical research. Several Indian surgeons have worked in his department and are holding senior positions in India.

The Bio-Data given below will attest to his prodigious output devoted to surgical correction of birth defects in children.

T. DORAIRAJAN

BIO-DATA OF PROF. F. DOUGLAS STEPHENS

Date of Birth	:	10-10-1913
Nationality	:	British - Australian
Current Position	:	Honorary Consultant Paediatric Surgeon, Royal Children's Hospital Honorary Research Fellow, Royal Children's Hospital Research Foundation
Year of Graduation	:	M.B.B.S. 1936 - University of Melbourne Post Graduate Degree Diploma - M.S. Melbourne 1943 FRACS Melbourne 1943

Medical Honours and Awards with dates :

1947 - 1950	Nuffield Travelling Fellowship to UK - Great Ormond Street Hospital for Sick Children
1949	Arris and Gale Professor, Royal College of Surgeons - UK.
1951	Nuffield Dominion Fellowship - Melbourne.
1962	Triennial Research Prize - BMA Victoria, Australia.
1964	Harry Swift Medal - Adelaide Children's Hospital.
1968	Fulbright-Hays Lecturer - (Australian-American Education Foundation) in USA.
1976	Denis Browne Medal - British Association of Paediatric Surgeons.
1986	The Urology Section Medal of the American Academy of Paediatrics.
1989	W E Ladd Medal of the Surgical Section of the American Academy of Paediatrics.

Honorary Fellowships or Memberships of other Colleges, Societies etc.

1968	Hon. Surgical Fellow, American Academy of Paediatrics
1971	Honorary Member, Australasian Society of Nephrology
1971	Honorary Member of the Argentinian & Brazilian Society of Paediatric Surgeons
1973	Honorary Member, Society Paediatric Urology
1976	Honorary Member, Urological Society of Australasia
1976	Honorary Member, Northwest Urological Society of America
1977	Honorary Member, Canadian Association of Paediatric Surgeons
1978	Elected Corresponding Member, American Association of Genito-Urinary Surgeons

1978	Elected to Honorary Membership, Australian College of Paediatrics
1981	Honorary Member, North Central Section of the American Urological Association
1981	Honorary Member, Association of Surgeons in India
1987	Honorary Member, British Association of Paediatric Surgeons
1988	Elected to Fellowship of Urological Society of Australasia
1989	Honorary Life Membership of the Australian Association of Paediatric Surgeons

Research Activities :

1958 - 1975	Director Department of Surgical Research, Royal Children's Hospital
1982 - 1986	Director of Research and Education, Children's Memorial Hospital, Chicago, USA

Research Appointments :

1947 - 1950	Research Fellow - The Hospital for Sick Children, Great Ormond Street, London, UK
1951 - 1957	Research Surgeon - Royal Children's Hospital, Melbourne, Victoria, Australia.
1958 - 1975	Director Department of Surgical Research - Royal Children's Hospital, Melbourne, Victoria, Australia.
1975	Director of Surgical Research and Education - The Children's Memorial Hospital, Chicago, Illinois, USA

Senior Office Bearer Appointments held in R.A.C.S., Universities and Charitable Organisations etc with dates :

1960	(approx) Chairman of Medical Staff, Children's Hospital, Melbourne.
1965 - 1976	Councillor of Royal Australasian College of Surgeons
1969 - 1974	Honorary Treasurer of Royal Australasian College of Surgeons
1969 - 1975	Editorial Secretary, Journal of Royal Australasian College of Surgeons (Australian & New Zealand Journal of Surgery)
1989	Chairman of the Archives Commitee, Royal Australasian College of Surgeons

Membership of Medical Association :

1937	Australian Medical Association
1942	Royal Australasian College of Surgeons
1948	Royal Society of Medicine
1953	British Association of Paediatric Surgeons
1968	American Academy of Paediatrics
1975	Society of Paediatric Urology of America (Honorary)
1975	Illinois Paediatric Surgical Aasociation (Honorary)
1978	Chicago Urological Association (Honorary)
1978	Corresponding Member of the American Association of Genito-Urinary Surgeons

Guest Lectures	:	26
Lectures given at National and International Meetings attended	:	23

BIBLIOGRAPHY - Books by F. Douglas Stephens :

1. STEPHENS, F D	:	Congenital Malformations of the Rectum, Anus and Genito-Urinary Tracts. E S Livingstone, London & Edinburgh, 1963
2. STEPHENS, F D & SMITH, E D	:	Anorectal Malformations in Children. Year Book Medical Publishers, Chicago, 1971.
3. STEPHENS, F D	:	Congenital Malformations of the Urinary Tract. New York, Praeger, 1983.
4. STEPHENS, F D & SMITH, E D (Editors) and PAUL N W (Associated Editors)	:	Anorectal Malformations in Children-Update 1988. Birth Defects : Original Article Series, Vol. 24, Number 4, New York, Alan R Liss. Scientific Publishers, 1988.

Papers Published	:	100
Additional Papers published by Members of the Surgical Research unit, Royal Children's Hospital under direction of F. Douglas Stephens	:	66

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President, Indian Association of
Paediatric Surgeons, Bombay

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3. Classification of Anorectal Anomalies
   Dr. V. Kalidasan

4. Newborn with Anorectal Malformation - Examination and Investigation
   Dr. D. Meera Bai

5. Role of Radiology in Anorectal Anomalies
   Dr. S. Rajah

6. Management of Low Anomalies
   Dr. H.R. Shanbhogue

7. Urologic Problems (Abnormalities) in
   Anorectal Anomalies
   Dr. P. Kesavan

8. Vestibular Malformations
   Dr. K.E. Mammen, Dr. Vinod Prem Singh
   Dr. Sudipta Sen, Dr. Ninan Zachariah
   Dr. Jacob Chacko

9. A Persistent cloaca
   Dr. S. Vasanthakumar

10. Colon Preparation
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11. Mechanism of Anorectal Continence in the Normal and Measures to Preserve it in
    Anorectal Malformations
    Dr. T. Dorairajan

12. Anorectal Anomalies - A retrospective Study
    Dr. P.N. Nagendran, Dr. Hayavadana Rao,
    Dr. K. G. Vivekanandan

13. A Review of Recent Literature
    Dr. Ravikumar

Anorectal Malformations - Historical Aspects

Dr. VINOD KAPUR
Dr. DORAIRAJAN

The reader interested in a history of Anorectal Malformations, should consult the writings of WEBSTER (1971) SCHARLI (1978) and DEVRIES (1984) and (1988). RUDOLF MATAS' paper "The Surgical Treatment of Congenital Anorectal Malformations considered in the light of Modern Operative Procedures" in transactions of American Surgical Association, Vol. 15, P 453, 1987 is often referred to in the literature as a landmark contribution on the history of these group of malformations.

In this short essay, the major historical landmarks are indicated. Out of a workshop organised by Stephens and Smith emerged the "WINGSPREAD" classification as well as a new method of assessing continence. A further salutory result of the Workshop was the book "ANORECTAL MALFORMATIONS UPDATE 1988" jointly edited by Stephens and Smith.

Special mention is made in this historical Update article about the contributions of Stephens in the elucidation of Sphincter anatomy, classification and operative procedures based on structure and function. The problem of deficits in continence following reconstruction has spurred study on the physiological mechanisms of continence involving manometry and electro myography. Great advances in diagnosis like C.T. Scan, MRI and Ultrasound have brought in further refinements in diagnosis and management of Anorectal Malformations.

These malformations occur with considerable frequency and hence there are references to their management from Egyptian and Graeco-Roman writings of antiquity to more recent times. The first recorded attempt at surgery was by PAUL of Agena (A.D. 625-690) who recommended a blind cut in the perineum with a bistoury and subsequent dilatations with bougies to keep the track patent. This was the treatment for several centuries till 1783 when DUBOISE of France performed the first left Iliac colostomy for the anomaly.

A major development from the long era of blind perineal cut was the formal exploration by a 'T' shaped incision first described by AMUSSAT of Paris in 1835. He dissected free the blind pouch and sutured the opened pouch margins to the perineal skin, heralding the era of perineal proctoplasty. Chassagnac in 1853 recommended left Iliac colostomy at birth and later a perineal proctoplasty.

In 1860 BODENHAMMER published a book "A Practical Treatise on the Aetiology Pathology and Treatment of the Congenital malformations of the Rectum and Anus". The well-known LADD and GROSS classification is mainly a simplification of Bodenhammer's classification. Bodenhammer also gave a clear description of the posterior saggital approach from the coccyx to the scrotum or the labium majus.

In 1896 Hadra recommended for the first time two stage Abdomino-Perineal Proctoplasty.

Developments in the First half of Twentieth Century.

in 1930 WANGENSTEEN and RICE described invertography to identify the blind pouch and this is still in vogue in the x-ray studies in the newborn. LADD and GROSS in 1934 shortened BODENHAMMER's classification and reported on their own results. Their very unsatisfactory Types I-IV classification was in vogue for over thirty years till it was supplanted by PATRIDGE and GOUGH'S, High-Intermediate-Low classification and its further elucidation in the "Proposed International Classification of 1970."

Yet another land mark in the mid-Twentieth Century was JONATHAN RHOADS report on one stage Abdomino-Perineal Proctoplasty in the newborn in 1948.

Contributions of F. Douglas Stephens

During the two decades from 1950-1970 working at the surgical research Department of the Royal Children's Hospital, Melbourne, as its Director, F.D. Stephens made meticulous dissection, histological and macrosection studies of preserved specimens and by investigation of patients under his care, and brought out with convincing evidence the key role of puborectalis sling in providing continence. He introduced the concept of the P.C. line now aptly termed by Paediatric Surgeons, Stephens line, to indicate the top border of the puborectalis sling. His technique of isolating the fistula from the urethra is unexcelled in its accuracy as well as ensuring safety of the delicate parts involved. In a series of publications finally culminating in the monograph "Ano-Recta Malformations in Children" (1971) jointly authored with E. DURHAM SMITH, Stephens set Paediatric Surgeons on the pathway of scientific reconstructive surgery of these complex malformations. Along with VARMA, Stephens further elucidated Neuro muscular reflexes involved in anorectal continence. Continuing the work at Chicago after he retired from the Children's Hospital, Melbourne, Stephens along with Smith organised the workshop at Racine from May 25 to May 27 at the WINGSPREAD Convention Centre.

The Racine Workshop and the Wingspread classification

The Racine Workshop was held at the Wingspread Convention Centre, of the Johnson Foundation, Racine, Wisconsin, U.S.A., Racine is about 80 miles north of Chicago on the shores of Lake Michigan. The workshop was held at the Wingspread Convention Centre.

Wingspread, one of Frank Lloyd Wrights creations, was the home of the Johnson family who converted it into a Convention Centre offering the premises, its refectory and secretarial services for scientific meetings, seminars and workshops.

"A study group of Paediatric Surgeons with special interest in Anorectal Malformations was assembled to evolve a suitable simple classification in the light of recent concepts and criticisms. A "high-intermediate-low classification with major sub types was agreed upon for males and females". The above in the words of Stephens and Smith (1986) was the stated objective of the workshop. A report on the workshop has been published under the title "Classification, identification, and assessment of surgical treatment of anorectal anomalies". Stephens and Smith Pediatr. Surg. Int. (1986) I 200-205.

Twenty Paediatric Surgeons from Australia, Canada, Germany, India, Japan, Mexico, Switzerland, South Africa, United Kingdom and United States attended the workshop. On three days, there were individual presentations, group discussions and arguments lasting late into the evening and finally emerged what is now known as the Wingspread classification, a newly proposed post operative assessment, and acceptance of Japanese study group registration form for standardised use by all.

The presentations from the study group were subsequently edited and presented in a book form by STEPHENS and SMITH as the Anorectal Update 1988.

Manometry and Electro-Myography in Anorectal Continence.

Another major historical development has been the elucidation of the physiological mechanisms of anorectal continence and their quantification. The contributions of NIXON, SCHARLI and KIESEWETTER, VARMA and STEPHENS, HOLSCHNEIDER, and YOKOYAMA have to be consulted. Together they have brought a measure of quantification of assessment of continence, particularly after the revision procedures in the disabled child.

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VARMA KK STEPHENS FRDE — Neuromuscular reflexes of rectal continence, AUST NZ J SURG 41: 263, 1972.

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HOLSCHNEIDER AM — The problem of anorectal continence. PROG PEDIATR SURG 9:85-97, 1976

Anatomy of the Anorectum

Review of the basics

Dr. V. KALIDASAN
Dr. H. R. SHANBHOGUE

This discussion of the anatomy of the anorectum reviews certain basic facts that are familiar to all surgeons.

- The surgeon's definition of anal canal is the part which extends from the cranial margin of the contracted puborectalis muscle to the anal orifice.

- At the pectinate Line :

i) skin of the anus changes to mucosa of the rectum;
ii) the level of the deep part of the external sphincter is the pectinate line;
iii) here is the lower limit of the puborectalis sling;
iv) it is the junction of the upper one-third and lower two-third of the internal sphincter;

-At the ano-rectal ring the epithelium becomes typical of rectal mucosa exhibiting crypts, goblet cells and mucus-secreting glands.

-The 'tail' muscles of animals are adapted to form the pelvic diaphragm and rectal sphincters of the human being;

-The pubo-rectalis, which is grouped with the levator ani is not found in lower animals;

-When the pubo-rectalis contracts it compresses the zone of anal canal cranial to the external and internal sphincters and effectively occludes the rectum, elongating the span of effective sphincteric action.

-The internal sphincter is a thickening of the circular muscle coat of the bowel; it is found chiefly distal to the pectinate line.

-The longitudinal muscle fibres 'the coat-tails' arise from the longitudinal muscle of the bowel wall and from the levator ani; these fibres penetrate the external sphincter muscles and are attached to the puckered perianal skin; these fibres knit the sphincters; they cause an ectropion of the perianal skin during defaecation that keeps the skin clean during the passage of motion; they are also called the "conjoined longitudinal ligament".

-The essential features of the levator- external sphincter complex to be remembered are;

The lowest and most medial fibres of the levator have a sling arrangement extending from the pubis anteriorly back to and behind the rectum and forward again to the pubis;

The levator supports the bowel and forms the floor of the pelvis;

The external sphincter is divided into strata from the skin up to the levator; the fibres are disposed both in a circular fashion and in the form of elliptical whorls attached anteriorly to the pubis or perineal body and posteriorly to the skin or coccyx.

It is not possible to accurately delineate between the lowest fibres of the puborectalis and most cranial fibres of the external sphincter.

It is important to note that in cases of supralevator anomalies the extent to which the external sphincter complex is developed and its arrangement shows considerable variation. This has to be borne in mind to reconcile the apparently different views of Stephens and Pena and DeVries; according to the former there being no external sphincter worthy of mentioning, and according to the latter there being a good strata of external sphincter muscles demonstrable during posterior saggital anorectoplasty.

Neurological aspects :

-Segments 2, 3 and 4 of the sacral spinal cord are the centres of arcs which subserve the receptors and effectors of the anorectum; these segments also subserve cutaneous sensation in the anal skin and perianal region;

The parasympathetic fibres arise from the anterior divisions of S 2,3 and 4; the preganglionic fibres unite to form the 2 nervi erigentes which go to the pelvic plexuses from where they are distributed to the pelvic viscera directly or via blood vessels; in the rectum they relay in the intramural ganglia.

-The nervi erigentes run lateral to the rectum in the Waldeyer's fascia close to the ischial spine or in the newborn at the level of the P-C line; the nerves can be safely separated from the rectum because of the natural plane of cleavage between the Waldeyer's fascia and the visceral layer of pelvic fascia investing the rectum; however, in anorectal malformations the rectum may not descend up to the P-C line and then these nerves run more medially in the Waldeyer's fascia below the rectum to reach the bladder base and in this situation they are vulnerable to injury-thus the importance of staying strictly in the midline during the posterior sagittal approach.

-The sympathetic nerves come from lumbar segments 2,3 and 4 unite to form the hypogastric plexus; they then continue as the presacral nerves to the pelvic ganglion which lies closely applied to the bladder base; this can be separated from the rectum as it lies in the parietal layer of pelvic fascia.

-The pudendal nerve, from anterior division of sacral segments 2, 3 and 4; it runs in the lateral wall of pelvis to supply the levator ani group.

-The perineal branch of the 4th sacral nerve supplies the external sphincter; the external sphincter is also supplied by the branches of the pudendal nerve.

-The arteries of the rectum and anal canal are:

i) Superior rectal artery being the continuation of the inferior mesenteric artery- this is the chief artery of the rectum- it divides into two branches which run on either side of the rectum, pierce the muscle coat and pass in the submucosa up to the anal columns;

ii) the middle rectal artery supplies the muscle coat of the lower rectum; it has poor anastomoses with the superior and inferior rectal arteries; it is a branch of the internal iliac artery (from the anterior trunk);

iii) the inferior rectal artery supplies the internal and external sphincters and the anal canal below the anal valves up to the perianal skin; it is a branch of the internal pudendal artery.

iv) the median sacral artery gives a branch to the posterior wall in the region of the anorectal junction.

The veins are :

i) the superior rectal veins commencing in the internal rectal plexus in the anal canal, run in the submucosa and continue as the inferior mesenteric vein;

ii) the middle rectal veins drain chiefly the muscular walls of the rectal ampulla;

iii) the inferior rectal veins begin in the external rectal plexus and drains the lower part of the anal canal.

The normal function of the anorectum and its musculature is discussed elsewhere.

PLEASE TURN OVER

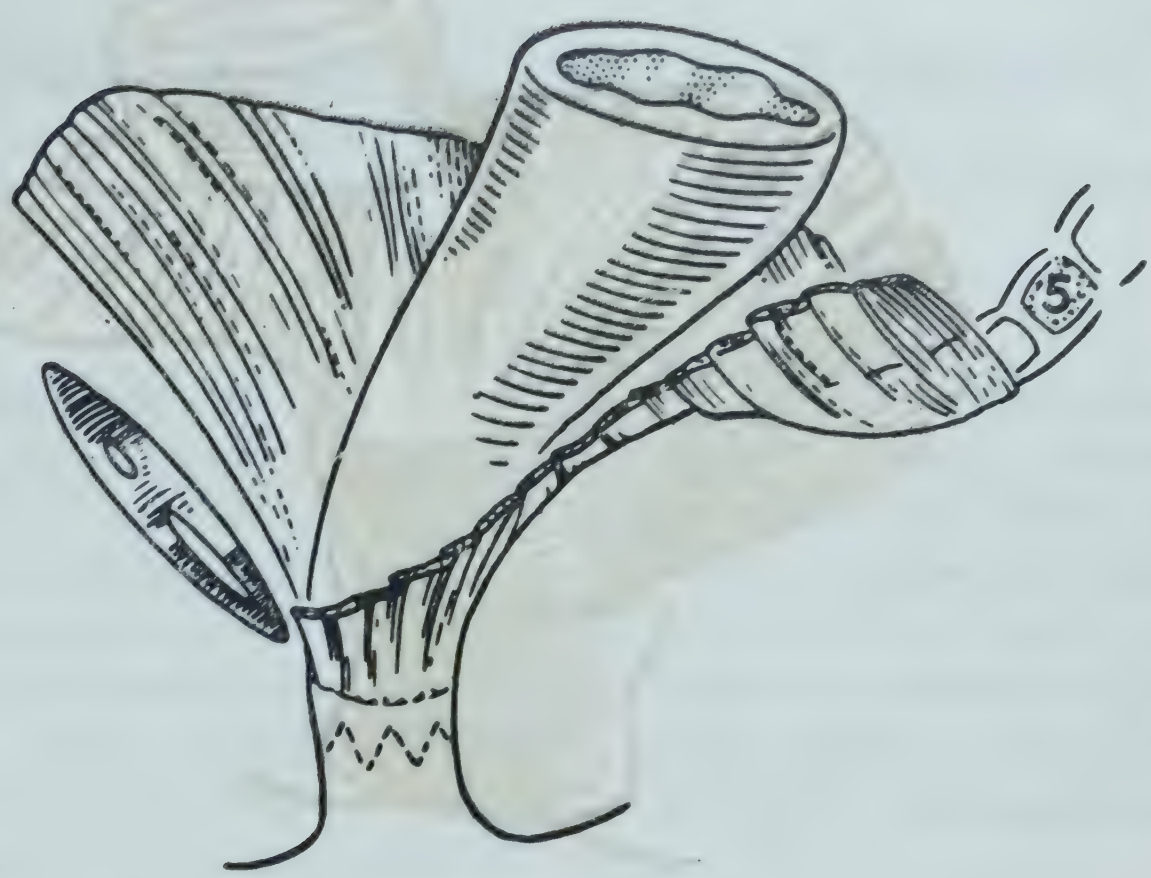
REFERENCES ;

1. Last R.J. : Anatomy, Regional and Applied, London, 1984. Churchill Livingstone pp. 328-330, 345-348.

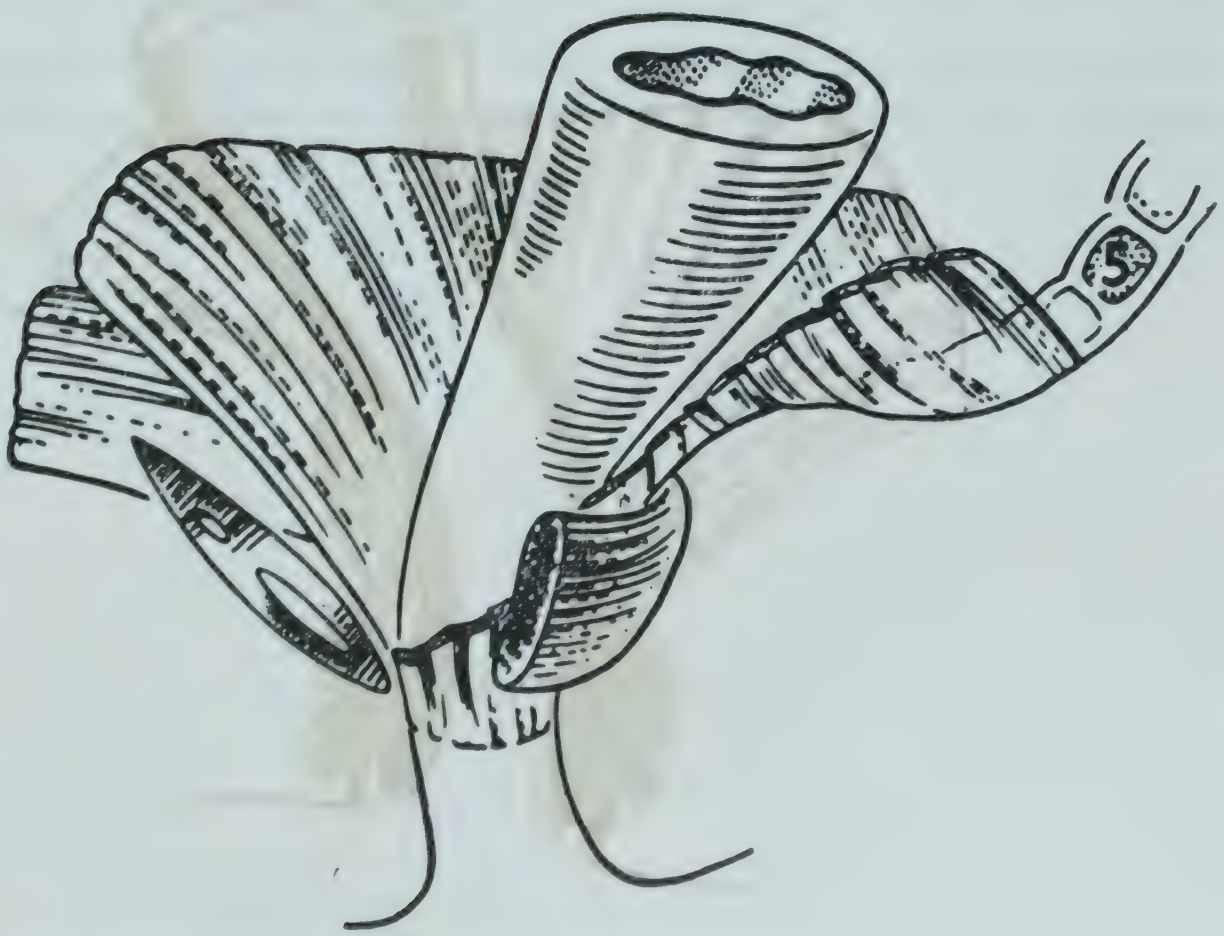
2. Stephens F.D., Smith E.D., : Anorectal malformations in children, Chicago, Year Book Medical Publishers, 1971.

3. De Vries P.A., Pena A : Posterior Sagittal Anorectoplasty, J. Pediatr. Surg. 17:638, 1982.

Acknowledgement : The adjoining figures are based on Ref. (2) above.

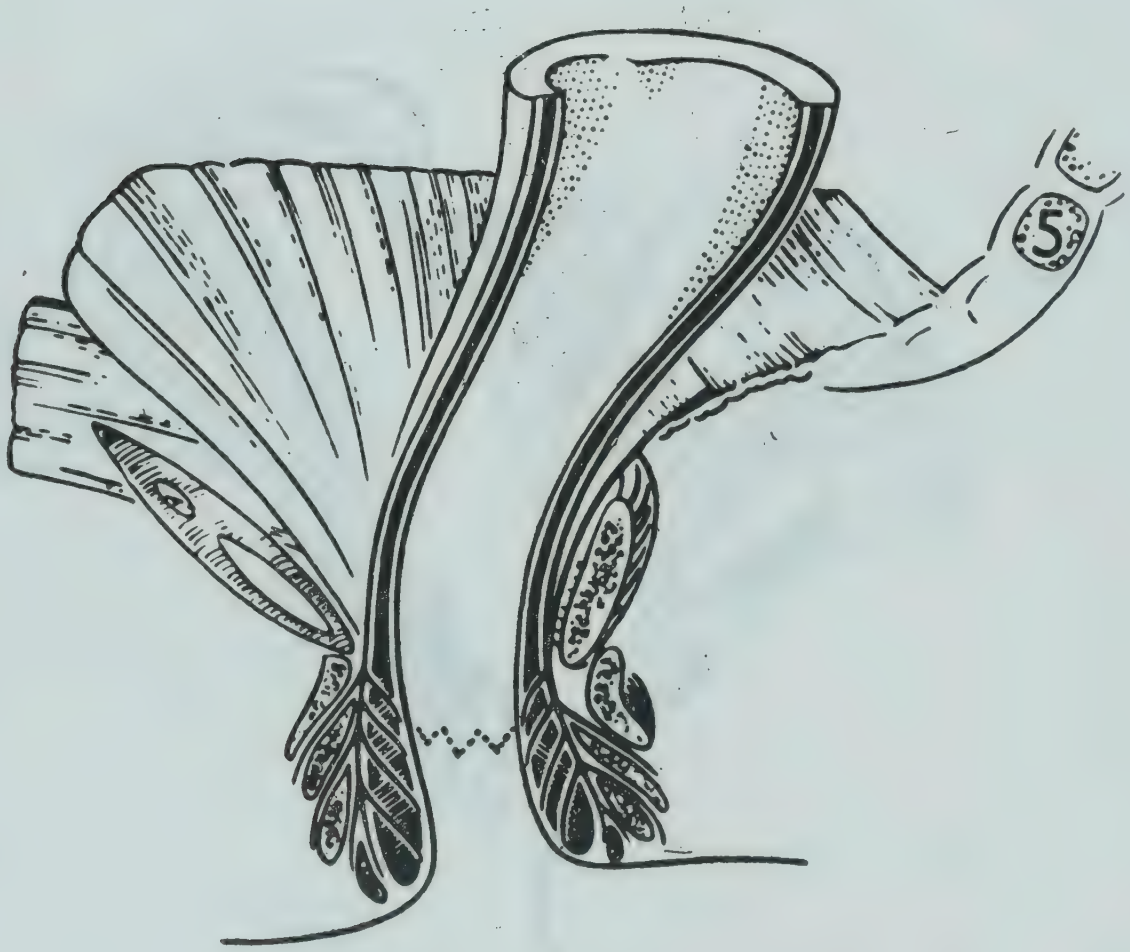


A. PUBOCOCCYGEAL SLEEVE



B. PUBORECTALIS SLING

C. PUBORECTALIS AND EXTERNAL SPHINCTER



D. CONJOINT LONGITUDINAL LIGAMENT

Classification of Anorectal Anomalies

V. KALIDASAN

We have come a long way from the days when all types of anorectal malformations were universally called "imperforate anus"; however, any attempt at classifying anorectal anomalies is bound to raise a hornet's nest of controversy. Stephens had organised a meeting that proposed a very detailed and complete international classification of these malformations (1970), familiar to all paediatric surgeons who are bound to have read the monumental book on anorectal malformations by Stephens and Smith (1971). The fact, however, was that to most people the classification was a little too arduous for day to day use. Many American surgeons still use the classification given by Ladd and Gross (1934) or its modifications (1980).

The most frequently used classification that has come into use over the last decade is the one popularly referred to as the "Wingspread classification" (1984). This was the result of a Workshop held at the Wingspread Convention Centre, Racine, Wisconsin between March 25-27, 1984. The importance of adopting a universal classification is to ensure that paediatric surgeons all over the world are referring to one and the same lesion when they compare various statistical data and analyse results of surgery. Failure to do this will result in people talking at cross purposes and interpretation of results will have no meaning. The table gives the Wingspread classification.

TABLE

The Wingspread Classification

LOW

FEMALE	MALE
Anovestibular fistula	Anocutaneous fistula
Anocutaneous fistula	Anal stenosis
Anal stenosis	

INTERMEDIATE

FEMALE	MALE
Rectovestibular	Rectobulbar-urethral fistula
Rectovaginal	Agenesis without fistula
Agenesis without fistula	

HIGH

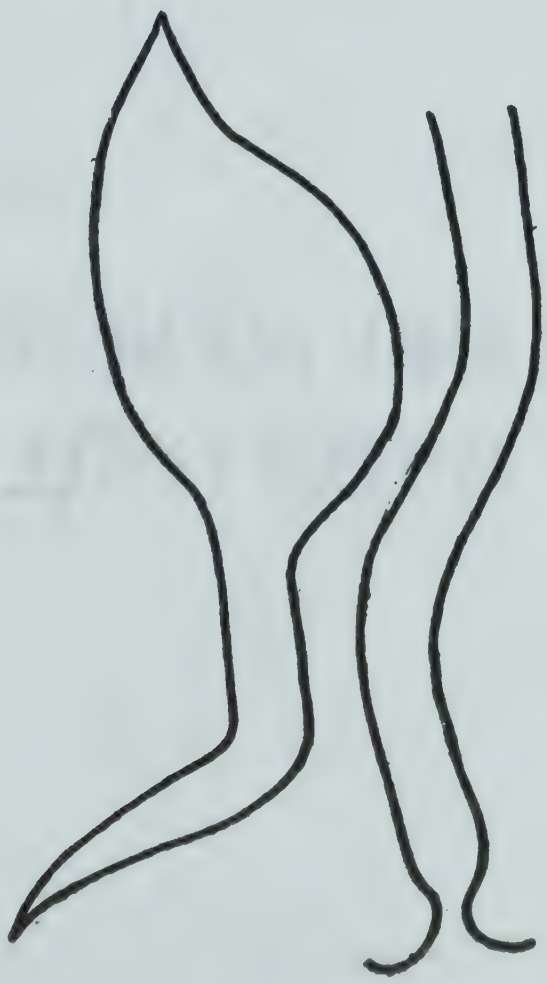
FEMALE	MALE
Anorectal Agenesis with Rectovaginal fistula	Anorectal Agenesis with Rectoprostatic-urethral fistula
without fistula	without fistula
Rectal Atresia	Rectal atresia

CLOACA AND RARE MALFORMATIONS

WING SPREAD

MALE
LOW ANOMALIES

1. ANOCUTANEOUS FISTULA.



2. ANAL STENOSIS.

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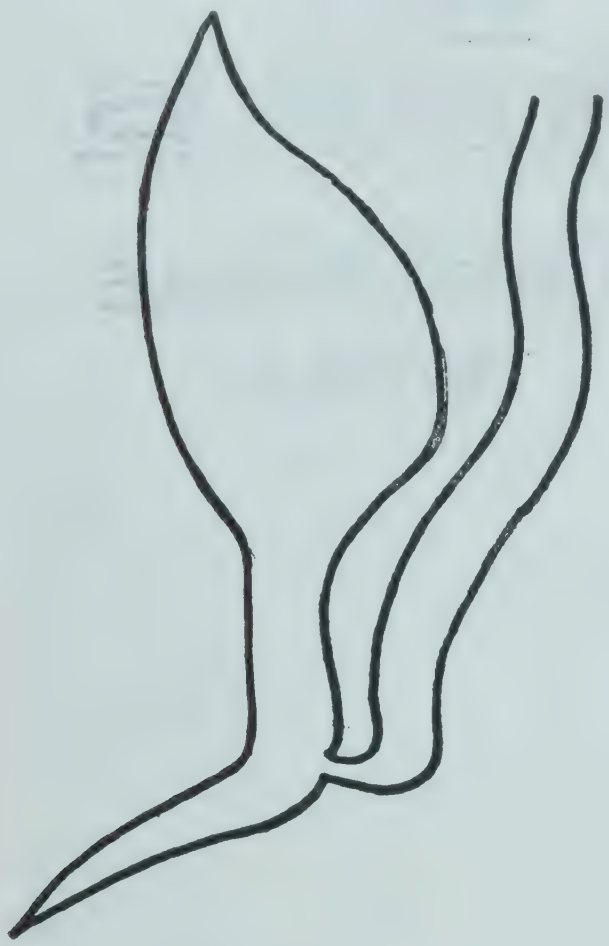
India

WINGSPREAD



MALE
INTERMEDIATE

1. RECTOBULBAR
   URETHRAL FISTULA

2. ANAL AGENESIS
   WITHOUT FISTULA.

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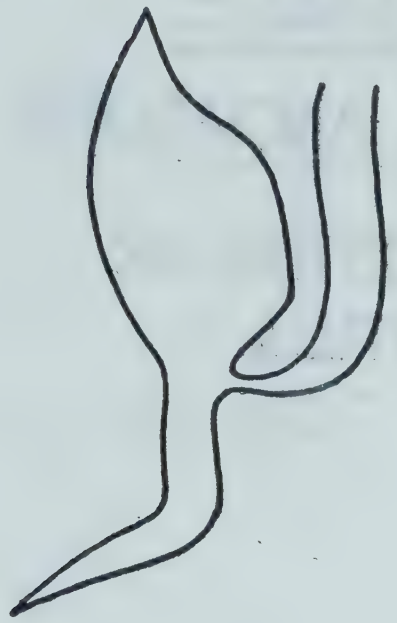
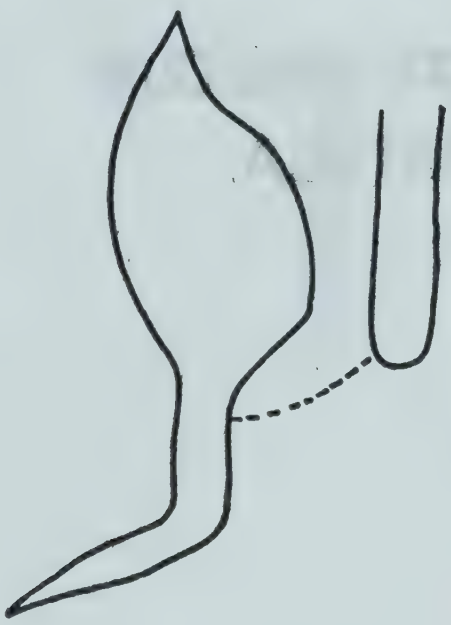
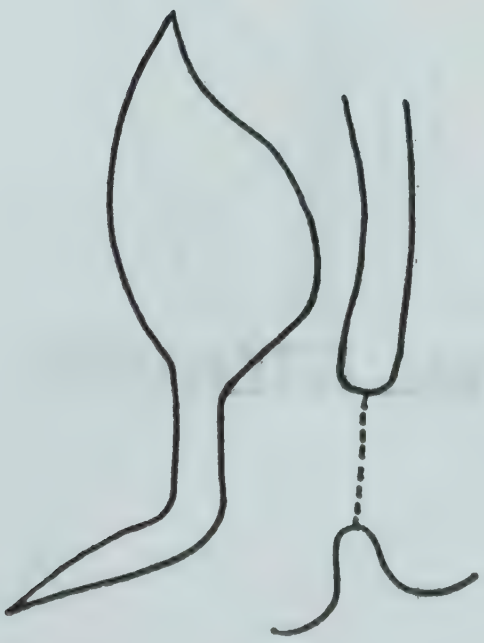


MALE
HIGH ANOMALIES

1. ANORECTAL AGENESIS WITH RECTO PROSTATIC URETHRAL FISTULA.



2. ANORECTAL AGENESIS WITHOUT FISTULA.



3. RECTAL ATRESIA.

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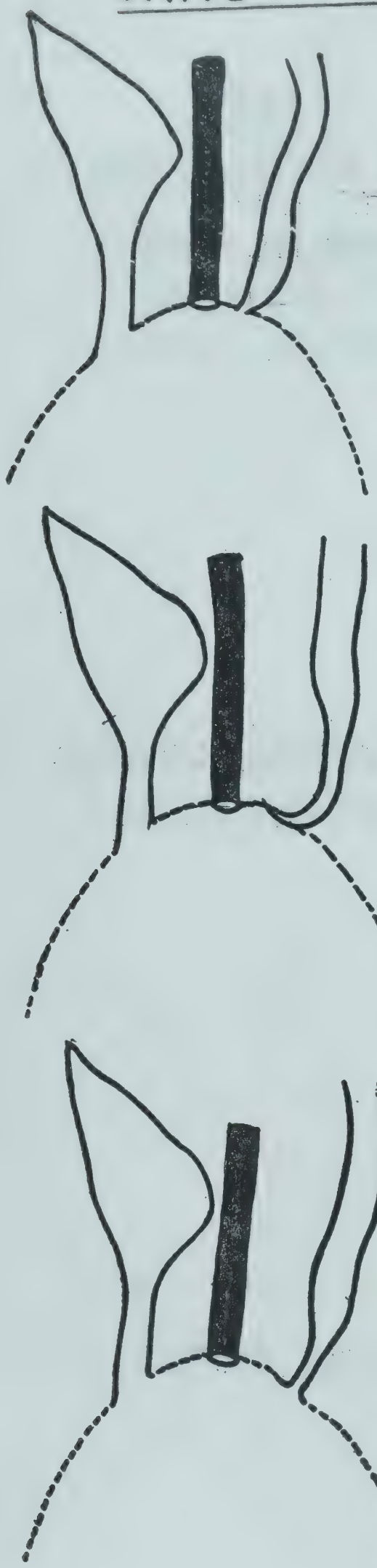


FEMALE

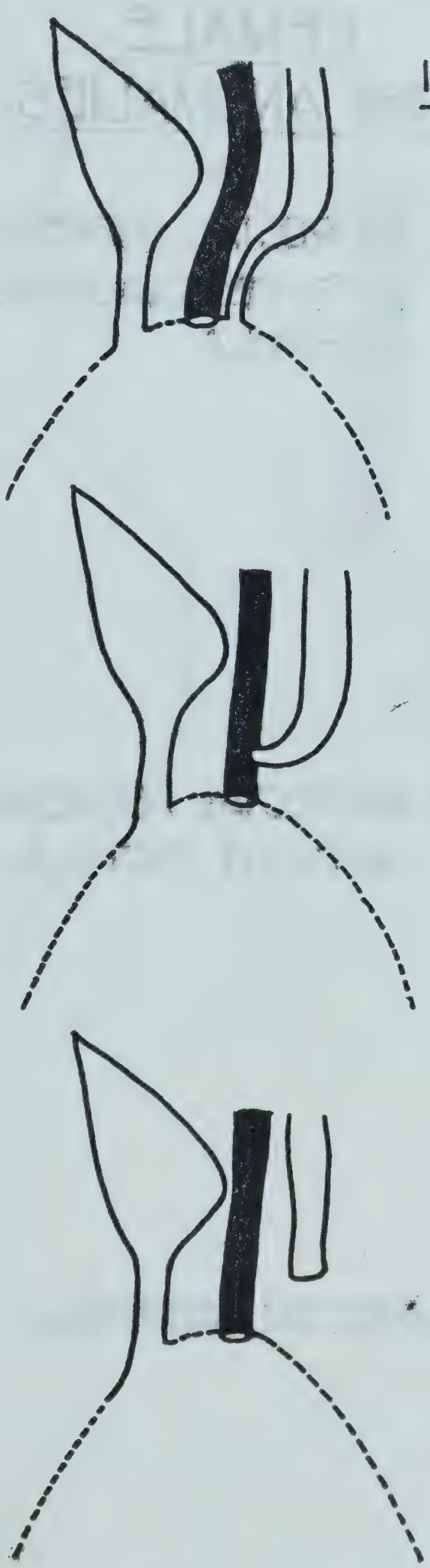
LOW ANOMALIES

1. ANOVESTIBULAR FISTULA.

2. ANOCUTANEOUS FISTULA.

3. ANAL STENOSIS.

WINGSPREAD



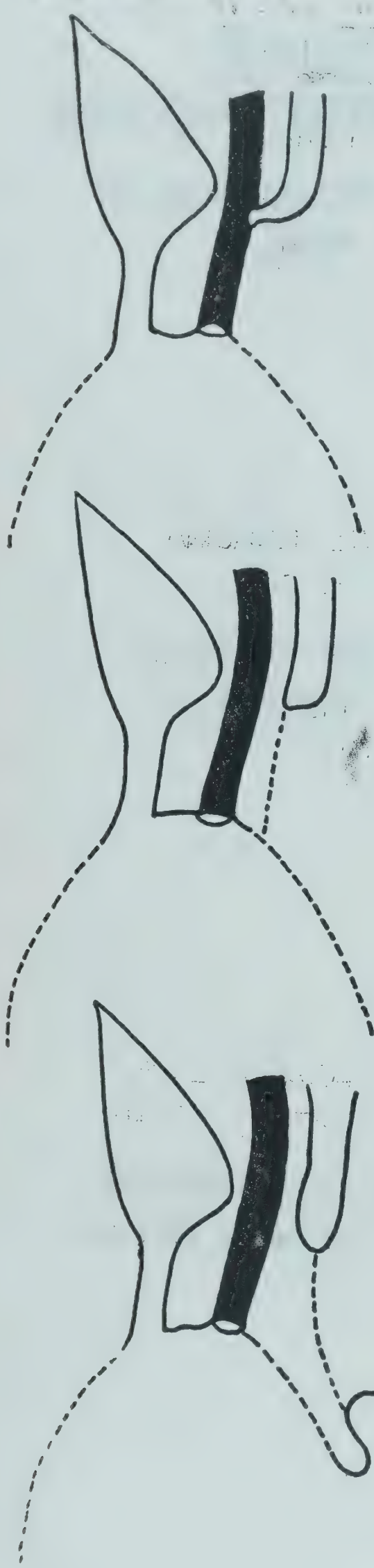
FEMALE
INTERMEDIATE ANOMALIES

1. RECTOVESTIBULAR FISTULA.

2. RECTOVAGINAL FISTULA.

3. ANAL AGENESIS WITHOUT FISTULA.

WINGSPREAD



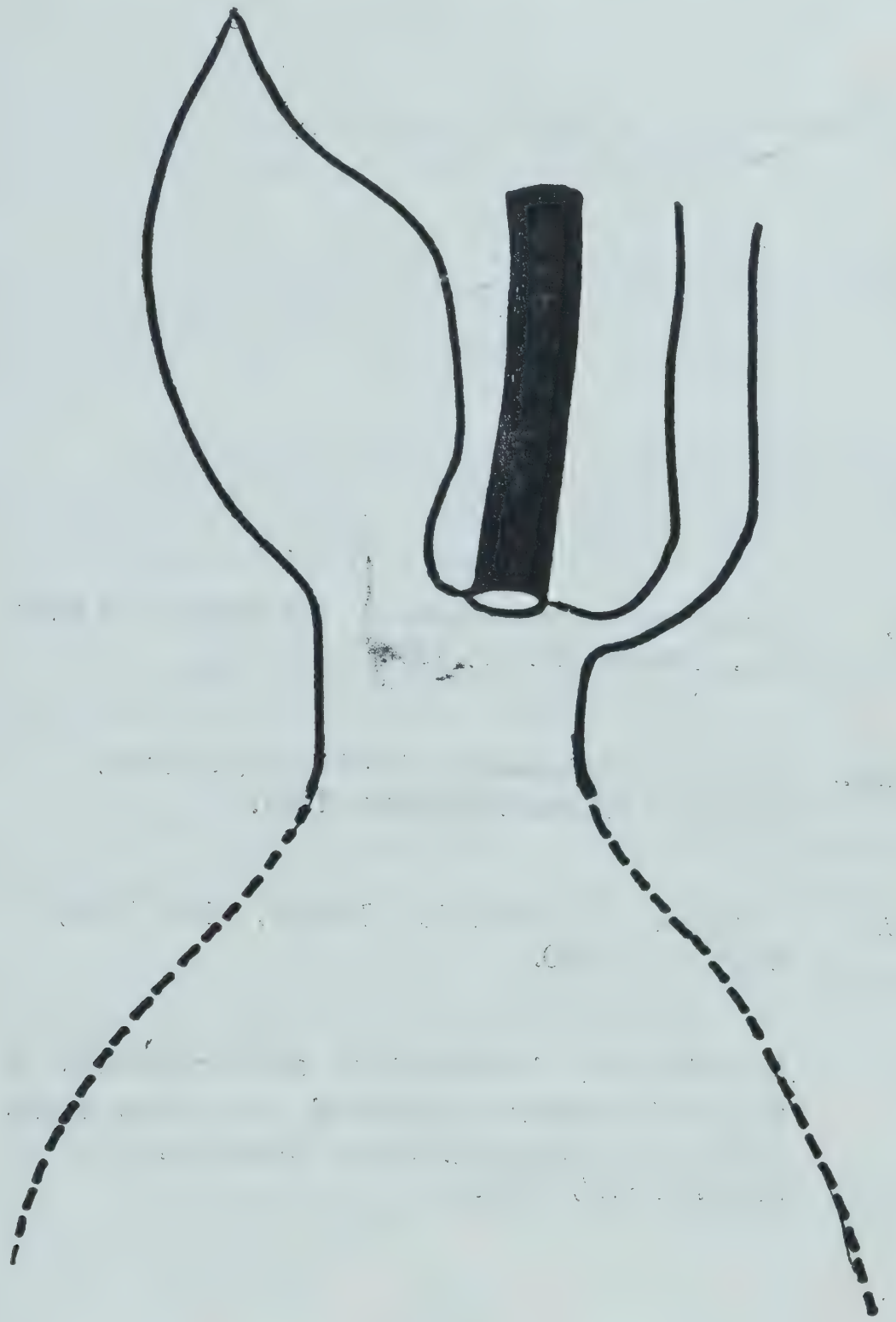
FEMALE
HIGH ANOMALIES

1. ANORECTAL AGENESIS WITH RECTOVAGINAL FISTULA.

2. ANORECTAL AGENESIS WITHOUT FISTULA.

3. RECTAL ATRESIA.

WINGSPREAD



CLOACA

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Newborn with Anorectal Malformation
Examination and Investigation

Dr. D. MEERA BAI

"Surgeon's Slogan should be Surgical Survivor Neither just Surgical skill nor Surgeon's Survival".

To achieve this goal in a newborn with ARM, accurate and early assessment of the (1) primary anomaly and (2) factors or secondary anomalies which may affect the surgical results adversely is essential.

In an ideal or sophisticated setup where the attending obstetrician detects the primary lesion early and the paediatrician picks up or excludes major life threatening anomalies or factors before passing the baby on to a surgeon, examination of the neonate by the surgeon centres round the local lesion.

However working knowledge of total evaluation is the crowning attribute of a surgeon and should not be dispensed with.

Majority of ARM present to the surgeon in neonatal period (65 to 75%); 25 to 35% newborns with ARM had associated anomalies while 25 to 30% succumbed in neonatal period with or without prompt surgery (personal study)

Some of associated anomalies encountered and aetiopathology of mortality in new borns studied are as shown in the tables.

Examination

It is ideal to commence with history. The important aspects which are of help to the surgeon in ARM are as shown in table IV

Physical examination
- General
- Regional
- Local

A. General Examination

Observation of colour, assessment of vital data and activity of the baby are important to exclude obvious and acute illness. Pink colour with normal heart rate and respiratory rate and Levocardia without cardiomegaly are important indices of normal function of Cardio-respiratory system. Deviation from these call for attention.

Lusty cry, brisk reflexes and vigorous activity (muscle tone) indicate normal CNS. Note that prematures and babies with Downs may be dull and limp.

B. Regional Examination

It is enough to (i) have a quick look for obvious malformations mentioned in table and to record them. Amongst these Downs, Potters facies, AMD syndrome or radial dysplasia are of importance as they may indicate occult and more serious anomalies of cardio-vascular or renal systems.

Whether there are obvious anomalies or not, further evaluation for internal anomalies is a must. OA with or without TOF is an important associated anomaly. It is clinicaly detected by noting the arrest of No. 6 or 7 rubber catheter passed orally into oesophgus (confirm with a sklagram). Duodenal or high jejunal atresia is suspected clinically by (1) aspiration of more than 20ml of bile from stomach in a baby 4-6 hrs old or noting the persistance of scaphoid abdomen after the age of 6-8 hours (confirm with erect babygram).
Congenital heart disease may be suspected clinically but confirmation by investigations is essential.

Disproportionately distended abdomen is equally important in ARM. It may be due to pneumoperitoneum, ascites or AMD syndrome.

C. Local Examination

(of Anogenital region)

Anorectal Malformation is a wide spectrum to-day; ranges from mermaid to microscopic anus.

It is neither a simple nor a single entity. It's complexity is due to

a) Intero-external extent of Anorectum
b) Close proximity to external genitalia
c) Bisexual occurence.

As a consequence clinical presentation and external appearances are variable and confusing and conflicting at times. Clarification needs careful assessment. The actual examination constituted by Inspection and Introduction of probe, dilator or digit depends on the presentation of the malformation.

Presentation of ARA

May be any one or combination of the following

a) Absence of external anal opening
   (i) with or (ii) without meconium track
b) Abnormal situation of anal opening
c) Abnormal size of anal opening
d) Normally sited anus
e) Failure to pass meconium (usually associated with a & d rarely with b or c)
   (given above)
f) Passage of meconium per
   - Urethra in male
   - Vagina in female
   - Abnormal orifice in both
g) Genital Ambiguity
h) Variation in number of orifices

Inspection

In lithotomy position with good light and relaxed and quite baby are rewarding. Gentle separation of labia in a female is required.

Inspect for any of the above presentations.

Introduction of probe. Catheter or dilator / digit

Absolutely indicated in female with external fistula.
Occasionally required in male.

In a female :

Introduce the probe into visible external orifice which is usually seen in vestibular fossa and note the direction and course of the probe. This helps to differentiate anovestibular from recto-vestibular fistula (probe passes posteriorly in the former and cranially in the latter)

In both male & female

a) Observe for arrest of probe introduced in a normally sited and sized anus arrest indicates Rectal Atresia

b) Assess the size of external orifice irrespective of its size to detect stenosis.

Depending on the findings of inspection and introduction of probe (commonly referred to as probing)

Categorise the malformation by alloting a name and frame fitting to the anomaly as given in table.

Investigations

Laboratory aids;

Are essential for assessment of associated factors like pallor, cyanosis and icterus or sepsis of newborn and not for ARA in an otherwise normal baby. In a baby with abnormal deviations, appropriate biochemical, bacteriological or hemalogical tests are indicated.

B Roentgenology / Imageology

The single most important investigation is conventional radiography. Radiological study is indispensible. Surgeon sans Roentgen in my view has no existance as an object without a shadow.

Invertogram

Without radio-opaque object convential skiagram to-day known as Invertogram is used to aid the surgeon to determine the level of atresia in ARA. It has limitations. It is unnecessary in some clinically recognisable low malformations. Useless when there is external or internal fistual while fallacious in babies less than 10 or 12 hours old. It is used in properly indicated cases which are few.

Erect Babygram with radio-opaque N. G. tube in site is found to be highly useful irrespective of age of baby in hours and type of anomaly. It is useful in detecting associated anomalies of heart, alimentary tract (OA, DA & IA or short colon) or respiratory disease or G. I. perforation which may adversely influence the ultimate outcome irrespective of class of anomaly or magnitude of surgery.

Amongst recent modalities of imageology, ultrasound CT and MRI are being tried to assess the extent of primary anomaly and its relation to puborectalis in some institutions. They may be of value if the technology is available and technician is well versed and reliable.

Summary and conclusions:

1) Emphasis in examination in new born with ARM is essential as majority of cases are seen in that period.

2) In the examination, equal emphasis is essential in general, local and systemic examinations in addition to history as neonate is not just a miniature adult and methods of assessment are different in babies.

3) In the examination and investigations detection of associated malformation and adverse factors are highly important as indices of prognosis and mortality, which the surgeon may have to face on one hand and communicate to the parents on the other.

4) In assessing the local anomaly simple clinical methods like inspection and or probing may be sufficient with appropriate use of a skiagram.

"Let our nation's baby boom
Not nipped in bud before full bloom
Let the hand that heals with knife
Hold and sustain baby's life"

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3) Case records studied over 25 years (author's cases)

Table I Obvious external anomalies

Head & Neck	Occular	Agenesis	2
	Aural	Ext Ear	3
	Oral	CL & CP	4
Face	Downs		16
	Potters		3 (1 still born)
Trunk	Myelomeningocele		4
	A. M. D. Syndrome		4
	Vesical Exstrophy		2
	V. I. F.		16
Extremities	Digits & feet (CTEV)		25
	Phocomelia		2
	Radial Dysplasia		4
Pernieum & Genitalia	Hypospadias etc		8
	Others		10
	Sacrococcygeal teratoma		1

Table II Internal malformations

Congenital heart disease	25
Oesophageal Atresia with or without TOF	10
Duodenal atresia	4
Oesophageal + duodenal Atresia	4
Ileo-jejunal atresia	2
Maltrotation	4
Double colon	2
Others	1

Table III Causes of mortality

Undetermined	34	
Respiratory & LBW & prematurity	14	
Tetanus & Temp derangement	10	
Haemorrhagic disease	7	
Sepsis of newborn	20	(pre - op 16)
Aspiration of vomitus	5	
AMD syndrome & G. I perforation	8	
Downs	11	
Cardiac disease	15	
Asso. Atresia of G. I. tract	22	
Lesion of urinary tract	3	

Table IV History

Period	Aspect	Its Value in examination
Prenatal	Infection Polyhydramnios	Look for Sepsis May indicate G. I. tract atresia (OA or DA)
	Oligohydramnios	May indicate renal or Urinary tract anomaly
Intranatal	Maternal fever Delayed labour etc	Look for sepsis
Postnatal	Apgar score Birth weight	Is of prognostic value Essential for Watertons classification.

Table V

Observation / Physical feature	Name	Frame	Sex
Absence of anal orifice			
a) with meconium track	Anocutaneous fistula	Low	Both
	Agenesis without fistula	Intermediate	Both
b) without meconium track	Anorectal agenesis without fistula	High	F/Both
Normally sited anus with arrest of catheter or digit	Rectal Atresia	High	Both
Abnormally sited external orifice In vestibular fossa	Ano Vestibular Fistula	Low	F
	Rectovestibular Fistula	Intermediate	F
Abnormal size (stenosed)	Anal stenosis	Low	Both
Passage of meconium per urethra	Rectobulbar Fistula	Intermediate	M
	Recto-prostatic Urethral Fistula	High	M
Per vagina	Recto vaginal Fistula	Intermediate	F
	AR Agenesis with RVF	High	F
Variation in number of orifices			F
Single common opening-cloaca More than 4 openings-Duplication			F

Role of Radiology in Anorectal Anomalies

S. RAJAH

INTRODUCTION

With tremendous technological advances modern radiology helps not only in diagnosis of the babies born with anorectal anomalies but also helps in planning the management and predicting the prognosis. Properly performed and carefully interpreted radiological investigations can give useful information regarding the decision to do a simple perineal operation or colostomy and even about the site of colostomy, a left Iliac or Transverse Colostomy.

AIMS OF RADIOLOGICAL INVESTIGATIONS

1) To determine the level and anatomy of anomaly
2) To identify any associated fistulous communications
3) To determine the presence or absence of any other congenital anomalies
4) The integrity of perineal nerve supply (Sacral anomalies), Spinal cord anomalies
5) The sphincter muscle complex (SMC) available for reconstruction.

RADIOLOGICAL MODALITIES AVAILABLE (Radiologist's Armamentarium)

I. X-ray

a) Plain x-ray chest, abdomen-supine
   Invertogram or prone lateral x-rays
b) Contrast study:
   1) Direct injection of radioopaque dye into the fistulous tract
   2) Percutaneously into the rectal pouch (Muragasu)
c) 1. Voiding cystourethrography
   2. Ascending urethrography
d) Intravenous pyelography

II. Ultrasound scan

III. Computarised Axial Tomography (CAT)

IV. Magnetic resonance Imaging (MRI)

Plain x-ray chest, abdomen and pelvis can help to diagnose associated anomalies like

1) Vertebral anomalies
2) Sacral anomalies
3) Cardiac anomalies
4) Oesophageal atresia with or without fistula
5) Duodenal Atresia and other bowel atresias like small bowel or colonic atresia
6) Cystic termination of the gut
7) Complications like spontaneous bowel perforation due to volvulus of sigmoid (distension and gangrene).

Iatrogenic perforation due to perineal exploration in a high anomaly.

Invertogram will not be useful when the bowel gas shadow does not reach the distal pouch in situations like oesophageal Atresia, Duodenal Atresia or small bowel atresias. Overdistended small bowel shadows also can give a deceptive picture in the invertogram.

INVERTOGRAM

Accurate interpretation of invertogram of the pelvis requires some thought and planning. The important precauticns to be taken while doing an invertogram are:

1) Invertogram should be taken after 12 to 18 hours of life to allow enough time for adequate bowel gas to pass to the end of the blind rectum. If the bowel is not overly distended with air one may wait 4-6 hours before placing a nasogastric tube for gastric decompression that will permit enough air to pass down to the gastrointestinal tract. But one should guard against overdistension of bowels because unrelieved massive bowel distension producing sigmoid volvulus and spontaneous bowel perforation can occur in some babies by 24 hours of life.

POSITIONING OF THE BABY FOR INVERTOGRAM

1) Baby should be held vertically upside down for 3 minutes before the film is taken
2) The baby should be quite and should not cry or move during exposure
3) The hips should be slightly extended or kept relatively straight so that the femora do not obscure the pubic bone
4) It is very important to obtain a true lateral view of the pelvis where both the right and left Ischium overlie each other exactly
5) Accurate centering on the greater trochanter
6) Placing a thin smear of barium paste in the buttocks cleft at the level of the external sphincter helps to denote the cutaneous level of the anus.

PRONE TRANSLATERAL X-RAYS

1) The baby is placed in Prone Jack-knife position for a few minutes
2) A lateral film is taken centering on the greater trochanter

ADVANTAGES OF PRONE LATERAL X-RAYS

1) Easy to perform and gives superior radiographs
2) Baby is not disturbed much during the procedure so baby can rest quietly in this position for a longer period
3) The chances of aspiration during the procedure is less especially in babies with tracheo-oesophageal fistulae
4) No need for anybody holding the baby in this position.

INTERPRETATION OF INVERTOGRAM AND PRONE LATERAL X-RAYS

This is based on the relationship of the air in the distal blind pouch to the pubococcygeal line and the 'I' point.

Pubococcygeal line is the line drawn from the upper border of the symphysis pubis to the sacrococcygeal junction.

P. POINT The centre of the 'boomerang' shape of the Os Pubis
C. POINT Is just caudal to the last (fifth) ossfic centre of sacrum

Pubococcygeal line cuts through the upper crescenteric margin of the ossified ischium i.e. the cranial one quarter with caudal three quarters of the Ischial shadow.

PUBOCOCCYGEAL LINE IN BABIES WITH SACRAL AGENESIS :

Can be developed by projection from the pubis through the same site on the Ischium which serves as a reliable alternative landmark. In these circumstances the P.C. line lies well caudal to the last ossified vertebra of the defective sacrum.

ISCHIAL LINE (I LINE)

'I' line is drawn through the 'I' point parallel to the pubococcygeal line
'I' point is the inferior end of ischial comma

STRUCTURES LYING AT THE LEVEL OF PUBOCOCCYGEAL LINE

IN MALE
1) Bladder neck
2) Veru montanum
3) Anterior peritoneal reflection of the rectum

IN FEMALE External OS of the cervix

STRUCTURES LYING AT THE LEVEL OF ISCHIAL LINE

IN MALE: Bulb of urethra

The urethral orifice lies caudal to the 'I' point. In Anal agenesis and in rectourethral fistula the gas shadow reaches the Ischial line and this is the lowest point of the levetor in these deformities.

IN FEMALE The 'I' line corresponds to the upper limit of the perineal body and the level of triangular ligament.

INTERPRETATION OF INVERTOGRAM

1. HIGH OR SUPRA LEVETOR ANOMALY :

The blind pouch ends at or above the pubococcygeal line

INTERMEDIATE : Air in the rectum ends between pubococcygeal line and Ischial point:

LOW OR TRANSLEVETOR ANOMOLY : Air in the blind pouch below the Ischial line

PROBLEMS IN INTERPRETATION OF INVERTOGRAM :

1) Gas shadow much higher than expected: May be due to contraction of the puborectalis muscle while taking pictures

2) Gas shadow is not smooth and rounded :

Due to active contraction of the puborectalis or meconium in the distal rectum. Holding the baby in the inverted position may succeed in displacing the meconium

3) Gas shadow lower than expected : May be due to the child straining excessively or due to excessive pressure on the abdomen.

Some intermediate lesions will appear to have a gas shadow below the 'I' point; occasionally in a male child with an intermediate anomaly with rectobulbar fistula the pouch may be filled with gas and lie below the 'I' point thus simulating a translevetor anomaly.

In female babies with vaginal fistula the invertogram may not reveal the level of blind pouch because of escape of gas and meconium through the fistula.

Presence of a 'Beak' anteriorly may identify the level of the fistula. But the 'Beak' may be present in some cases even without fistula.

Gas in the vagina or a low lying loop of small bowel may mimick a low lesion.

AIR IN THE BIADDER :

Invertogram may show air in the bladder which in a female indicates rectovesical fistula and in a male may be rectovesical or rectourethral fistula.

CONTRAST STUDY :

1) Percutaneous injection of soluble contrast material through the perinum into the distal pouch may be useful in demonstrating the distal end of the bowel and in outlining a fistulous connection to the genito-urinary tract.

2) Direct injection of a fistula or anus :
   a) Useful in defining the level of an anorectal stenosis, anovestibular or rectovestibular fistula
   b) Useful in distinguishing between rectobulbar and rectourethral fistula.

This information can be obtained from voiding cystourethrography or retrograde urethrography.

Micturition cystourethrogram : This is performed at an early stage which may find the site if any of a rectourethral or anourethral fistula. Even if the fistula is not visualised there is usually some characteristic angulation or tenting at the site of the fistula or a telltale streak from the urethra directed posteriorly near the ischial spine.

It may document an associated urinary abnormality especially vesico-ureteric reflux.

Can demonstrate rare deformities like rectourethral fistula without rectal or anal agenesis and duplication of urethra.

INTRAVENOUS PYELOGRAPHY : It helps to assess the structure and function of the upper urinary tract. Very useful in demonstrating renal dysplasia or agenesis.

COLOGRAM : Generally performed before the child goes home, 2 to 3 week after operation or during the early follow up period around 2 to 3 months of life.

1) confirms the level of anomaly suspected by invertogram
2) may outline the fistula or may show a 'beak' at the site of fistula
3) can give valuable information about the length of the distal loop so that a crucial decision about need for combined abdominal approach can be planned in very high anomalies.

ULTRASOUND SCAN OF PELVIS :

Proves to be very valuable in identifying the associated genitourinary abnormalities

The level of the distal pouch can be accurately outlined by careful interpretation of ultrasound. Gas in the distal pouch and full bladder are prerequisite for a proper interpretation.

ADVANTAGES OF ULTRASOUND :

- very simple
- accurate
- available in almost all centres now
- no radiation

COMPUTERIZED AXIAL TOMOGRAPHY (CAT)

Is very useful in the initial assessment of neonates with anorectal anomaly and also can give helpful indications for or against 'Re-do' procedures.

CAT can help in demonstrating the site and development of pelvic musculature (sphincter muscle complex)

CAT is useful in identifying sacral and spinal abnormality, spina bifida occulta, sphincter muscle complex deficiency, (hypoplastic spincter muscle complex.)

The conventional axial views best demonstrate the levator sling and its relation to neo-rectal placement

Coronal views give a better estimate of the bulk of the sphincter muscle complex [SMC]

CAT helps to assess the state of SMC and to define the eccenteric position of the rectum in postoperative patients with problems.

CAT is a valuable preoperative modality and could fundamentally influence both the technique of surgery and the prognosis.

MAGNETIC RESONANCE IMAGING [MRI]

It is the newest diagnostic tool that is capable of generating images of sections of the body in any plane.

MRI scanner permits better resolution of soft tissues than CAT scan.

The anatomical relationship of the most distal portion of the bowel to the muscles of continence can be directly visualised with good definition. Impacted meconium in the distal pouch serves as an excellent contrast agent because of high lipid content. The fistula can be accurately identified, lipomas in the sacral spine and other bony abnormalities are well demonstrated and it is believed that they should be corrected before any reconstruction of the rectum (if they are present) so that optimal innervation of muscle will be preserved.

MRI will give important details regarding the length and caliber of the cloacal channel, level of confluence of the urinary, genital and intestinal tracts.

MRI is very useful in demonstrating lesions of the spinal cord such as tethered spinal cord or neoplasms, sacral agenesis and thoracolumbar spinal anomalies.

Genit urinary tract abnormalities are very clearly outlined by MRI.

MRI will be taking an important place in evaluating the level and other abnormalities associated with anorectal malformations in future when the technique is more readily available.

ACKNOWLEDGEMENT

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Management of Low Anomalies

DR. H. R. SHANBHOGUE

Management of these anomalies can be discussed under

I. Management of low anomalies in the male
II. Management of low anomalies in the female

IN THE MALE

I. Ano - cutaneous fistula (covered anus incomplete)
II. Anal stenosis - under this heading of stenotic lesions the following conditions are seen in the clinical practice :

a. Covered anal stenosis
b. Anal membrane stenosis
c. Imperforate anal membrane

Anocutaneous fistula :

This lesion is the most common of the low anomalies occuring in the male. In the common form the anal canal is of normal calibre and correctly located in the pelvis and perineum and is complete to the level of anal valves. From the terminal normal anal canal, the fistula runs forward and opens in the perineal raphe, anywhere adjacent to the anal site to the ventral surface of the penile shaft. Most commonly the opening is behind the scrotum. It is superficial & covered by thin skin. Meconium or epithelial plugs are seen at the orifice.

Careful inspection of the perineum is mandatory. Generally a fine orifice of fistula is found from the normal anal site to as far forward as frenulum of the penis. A speck of meconium will suggest the site of orifice. The fistulous tract is suggested by the bluish line in the perineal raphe or by the bulge of the meconium.

SPHINCTER:

There is plenty of controversy about normal sphincter availability to the caudal part of the anal canal. Proximal part of the anal canal and the rectum is invested with normal Levator complex. Distal part of the anal canal and fistula when present (though there is a controversy) generally it is agreed has an adequate internal and external sphincter complex. This has a great bearing on the outcome of the management, as the continence in this children following treatment is excellent.

Investigations like invertogram and fistulogram are unnecessary and can also be misleading.

Other anomalies like TEF, vertebral and sacral and renal should be ruled out.

Surgical management

Simple cutback procedure followed by post-operative dilatation.

Post-Operative care

1) Perineal cleansing of the anus with saline in the first week
2) Anal dilatation periodically
3) Prevention of constipation
4) Commencement of toilet training

Alternatively cutback followed by Y-V. Plasty has a lot to offer in preventing skin level stricture.

Anal Stenosis

a. Covered anal stenosis

This is the commonest form of anal stenosis. The genital folds may cover the anal orifice either anteriorly or posteriorly presenting the orifice stenotic.

Management : Incision of hypertrophic genital folds and minimal excision of the same and re-arrangement of perianal skin.

Regular post-operative dilatation.

Late trimmings of tags if necessary.

b. Anal membrane stenosis

This defect is rare.

Here the membrane is partially perforate.

This lesion sometimes is overlooked and the child presents later with chronic constipation, ribbon stools and secondary megacolon.

This condition is to be diagnosed from clinical appearance alone.

Invertography is unnecessary.

Management

Requires periodic dilatation.
Associated skin tags may be excised at a later date.

Imperforate anal membrane

This lesion is rare.

Anus is normally placed.

Perineum appears normal.

Condition presents as intestinal obstruction in the newborn.

Reveals a membrane 1 cm inside the anal verge.

On straining the membrane appears as a bluish bulge (due to meconium).

Invertography rarely necessary, when done the gas shadow is shown at the lowest level.

Treatment - Incision of the membrane followed by perineal anoplasty.

Anterior perineal anus

In wingspread classification it is included under anocutaneous fistula. In the update Stephens et al feel this form should be retained.

Here the anus is normal in appearance but is situated more anteriorly than normal position.
No treatment is necessary.

In small percentage of cases there may be constipation due to cul-de-sac, when a posterior incision is made to eliminate the cul-de-sac. Transplantation may be necessary if the shelf effect causes intermittent constipation.

Low Anomalies in the female

Ano vestibular fistula : This is the commonest lesion seen in the female child.
Clinical examination of the perineum shows three orifices in the vulval area (a) urethral (b) vaginal (c) bowel

A probe passed into the orifice passes backwards just behind the skin. This anomaly should be diagnosed by clinical examination alone.

Management : There is a controversy in the management. A simple cut back with dilatation of the orifice is a simple procedure and gives good results.

However the vestibular anus is aesthetically displeasing and many workers feel transplantation into the normal site is more appropriate. We agree with this view. Transplantation can be done at 3-6 months of age or with good weight. Ano cutaneous fistula and anterior perineal anus present and are managed just as in the male.

A stenotic orifice may be found at or near the normal anal site & the fistulous orifice may be found anywhere in the midline of the perineum between the normal anal site and the vestibule. The management depends on the choice between the simple cut back followed by dilatation and transplant. We favour the transplant of the anal opening to its normal site.

Anal stenosis in the female is similar to the male in presentation and management.

Perineal canal and imperforate anal membrane are operated by a perineal approach.

Perineal groove does not require surgery.

SUMMARY

All the low anomalies in both female and male children can be diagonised accurately by clinical examination of the perineum alone.

Invertogram is rarely necessary and it may be misleading.
Fistulogram may be necessary.

Levator complex is essentially normal in these lesions. Internal and external sphincter is adequate in these lesions. Hence the continence is excellent following surgery.

Local procedures like cutback, Y-V anoplasty and transplantation procedure are sufficient and major procedures like sacro perineal or posterior saggital recto-anoplasty are not necessary for this lesions.

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Urologic Problems (Abnormalities) in Anorectal Anomalies

Dr. P. KESAVAN

INTRODUCTION :

It is not difficult to understand that urologic problems are encountered often in dealing with Anorectal Anomalies. We all know that, a good number of the cases have a fistula in communication with the urinary tract like Anorectal Anomaly with Recto Urethral Fistula or ARA with Rectovesical Fistula etc. Naturally urinary infection and problems associated with its disconnection are encountered by the surgeon. Apart from these Urologic Problems inherent in this types of fistulous Anorectal Anomalies, the other urologic problems that we see in this disease could be caused by either an abnormal development of the urinary tract itself like duplication, mega ureter or reflux etc., or caused by the asscciated abnormalities of other systems (like sacral agenesis or myelomeningocele).

CAUSES :

So we may group the urologic problems in Anorectal Anomalies under 4 heads.

1) Those problems inherent in fistulous anomalies.
2) Those problems which are caused by their treatment - iatrogenic.
3) Those caused by the associated malformations of the urinary tract.
4) Those caused by the associated malformations of the other systems.

It is obvious that the groups 1,3 and 4 are developmental in origin and the group 2 is iatrogenic.

EMBRYOLOGICAL BASIS FOR DEVELOPMENTAL LESIONS :

We all know that cloaca is very intimately associated with the development of both the anorectum and the urinary tract in the fetus. It is the cloaca which gets divided into an anterior chamber called Urogenital Sinus (UGS) and a posterior chamber called Rectum by the formation of the Urorectal septum. We also know that it is into this UGS that the Mesonephric Ducts (MND) open. In its turn, it is these MND which give origin to the Ureteric Buds (UB). UB is the starting point of the formation of the ureter and kidney. So any abnormal or aberrant events in the further development of the Cloaca will result not only in abnormalities occurring in the Anorectum but also in the urinary tract depending on the time, site and duration of the influence of the teratogenic factor; the organ deranged will be genitalia, bladder, ureter or kidney. The occurrence of the associated abnormalities of the spine also could be based on the extended influence of the teratogenic factor at the tail end of the fetus; these also contribute largely to urinary problems in anorectal anomalies.

IATROGENIC CAUSES :

The second group of the urinary problems that we encounter in anorectal field are ones caused by the treatment. They are either A) Intraoperative injuries to urinary tract or postoperative complications eg., injury to bladder or ureter/infection, abscess forma-

tion and fistula developing postoperatively. or B) We may encounter in some, a residual anomaly left behind inadvertantly or by an inadequate operation, like a diverticulum in the posterior urethra (a residual stump of the fistula) causing recurrent urinary tract infection or obstruction to urinary flow and its consequences. Or C) Injury to the nerves which supply the bladder neck and the sphincter during dissection at and around the fistula.

MANIFESTATIONS OF THESE LESIONS :

The manifestations of the urinary problems in Anorectal Anomalies are generally in the form of either recurrent urinary tract infection, or dribbling. In a few neglected cases or severe involvement of the urinary tract in the maldevelopment, they present with renal failure. Oftener than not, the iatrogenic cases present for dysuria also, besides having dribbling or urinary leakage through anus.

It may be safely be said that, in general, the high Anorectal Anomalies have higher incidence of urinary problems, because 1) they have often fistulous communication with the urinary tract like rectourethtral or rectovesical fistulae and 2) they are also often asscciated with sacral agenesis or other spinal deformities.

Preoperatively, in these fistula cases, there is contamination of the urinary tract with faeces, causing urinary infection. This is possible and occurs even in those cases with by-pass like colostomy, unless appropriate and adequate cleaning of the distal bowel is carried out. The infection may become worse and may lead on to serious consequences, if it is not recognised and treated. Another cause for intractable and recurrent infection is the associated vesico ureteric reflux or megaureter. So in addition to treating urinary infection, one should always investigate to exclude these associated anomalies. Sacral agenesis is another condition that one should keep in mind. They manifest also for dribbling of urine.

Post operatively, persistent infection must put one on guard and investigations must be instituted to identify residual pouches of the rectum left attached to the urethra. This happens as a result of inadequate removal of the fistulous tract. Difficulty in passing urine may also be present. Dysuria may also result from injury to the urethra and subsequent development of a stricture urethra or a new fistula may develop into the anal canal resulting in urine leaking continously through the newly created anus.

It should be pointed here, that there are many children in whom a combination of the causes operate and the urinary problems are really intractable and some of them may go on to stage of renal failure.

There is another small group of children with Anorectal Anomaly, where the urologic abnormalities are a part and parcel of more complex syndromes like VATER SYNDROME. Various other systems are involved in this syndrome and some of them are life threatening in their own merit.

RECOGNITION AND MANAGEMENT :

This may be considered according to the causes described above.

In those patients of High Anorectal Anomalies with fistula, the recognition of urinary problems like urinary infection is dependent on a high degree of awareness on the part of the clinician and a close watch must be kept on these children. The best way to manage is, of course, to prevent urinary infection by giving an adequate way out to the distal bowel through colostomy stoma. This is better done immediately after colostomy, otherwise cleaning becomes really difficult and inadequate. Once infection is recognised it should be adequately treated and investigations of the urinary tract must be carried out like voiding Cystourethrography, Ultrasonagraphy and Intravenous Pyelography. If they reveal any other abnormality in the urinary tract like Urethral Reflux or Megaureter, then the treatment of the urinary infection must be prolonged and the disconnection of the fistula with anorectal corrective surgery may have to be carried out at a much earlier period than usual. Or alternatively when the child is not fit for a major anorectal surgery or when the urinary infection is really severe and not controllable, correction of the urinary anomaly may have to be resorted to even earlier than the definitive surgery of the anorectal anomaly. If there is renal failure or life threatening emergency, a diversion procedure like cutaneous ureterostomy also is resorted to. On the other hand, if the urinary anomalies are asymptomatic, they are treated on their own merits after the anorectal correction is completed. In principle, management of this group of urinary problems in Anorectal Anomaly is not any different from those without anorectal anomaly.

On the other hand, we must be concerned more on the other type of urinary problems in Anorectal Anomalies, namely the iatrogenic ones, i.e., those caused by an inadvertent or inadequate surgery. This group is more relevant for our workshop to day, because they are all preventable. A conscientious anorectal surgeon must give equal importance to both - to achieve a 'good anus' and also not to injure the urinary tract. When surgery is done for high anomalies, it is very important to resort to very stringent and accurate technical execution of the procedure. Some of the very simple technical aids available for this purpose are : 1) a simple gadget of muscle stimulator, 2) cystourethroscopy, 3) urethral catheterisation and 4) catheterisation of the fistula with the aid of scopy before surgery. By these simple aids one can achieve very satisfactory result of a 'good anus' as well as intact urinary tract.

Another iatrogenic cause is the damage done to the nerves which supply the bladder neck and sphincters by unnecessary and out of limits dissection in the area of the fistula. The resulting neurogenic bladder causes uncontrollable dribbling, dysuria and urinary infection. Similar symptoms occur with those cases associated with sacral agenesis also and are difficult to cure permanently and hence, the emphasis on prevention.

Inadequate removal of tne fistulous tract results in a small diverticulum like pouch attached to the posterior urethra / bladder, causing a nidus for infection. It may expand in size and cause obstruction to urinary flow. Sometimes, an abscess may form in that region and may break down to produce a fistulous track to the exterior or to anal tract resulting in a cutaneous or anal urinary fistula. These may be reoperated after careful evaluation. The outcome is variable ond oftener than not, they are disastrous.

CONCLUSIONS :

Though the urinary problems in Anorectal Anomalies can be considered under 4 different causative angles, they really fall under 2 broad groups - Developmental and iatrogenic.

The high anomalies are prone to get more of urinary problems, though even the low anomalies are associated with maldeveloped urinary tract.

Barring the fistulous cases, the urinary problems caused by the associated anomalies are treated on their own merits, mostly. Sometimes the definitive surgery for the ARA or the corrective surgery for the urinary anomaly may have to be preponed.

Much more important and relevant are the iatrogenic problems, because they are preventable. A good paediatric surgeon must be concerned equally with obtaining a 'good anus' as well as an intact urinary tract and its nerve supply.

VESTIBULAR MALFORMATIONS IN ANORECTAL ANOMALIES

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VESTIBULAR MALFORMATIONS IN ANORECTAL ANOMALIES

The simplified 1984 Wingspread Classification, groups the low type of anorectal anomalies with fistulous communication between the "Hind Gut" and the vestibule, the vulva and the perineum into one common group, as "Anovestibular Fistula". This group also includes vestibular, vulval and anterior perineal anus. The intermediate type of anal aganesis with rectovestibular fistula in the International Classification of 1970, is classified as"Rectovestibular Fistula" in the Wingspread Classification. The terminologies in the international classification is more precise than the simplified Wingspread Classification.

In this chapter, on Vestibular Malformations, all common as well as rare types of anorectal malformations in the female with fistulous or direct communication from the "Hind Gut" to the vestibule, vulva or perineum are dealt with.

Cloacal anomalies, rectovaginal fistula, anorectal agenesis without fistula and anal agenesis are excluded.

TABLE - 1

VESTIBULAR MALFORMATION - CLASSIFICATION	
I N T E R N A T I O N A L	**W I N G S P R E A D**
INTERMEDIATE	**INTERMEDIATE**
Anal Agenesis with Recto-Vestibular fistula	Rectovestibular fistula
L O W	**L O W**
(TRANSLEVATOR)	
Anterior Perineal anus Vulvar Anus Anovulvar fistula Anovestibular fistula	Anovestibular fistula
Miscellaneous Anomalies	**Rare Malformations**
Perineal Canal	Perineal Canal
Perineal groove	Perineal groove

Vestibular Malformations are the commonest type of congenital anorectal anomaly seen in the female child (seventy seven)

TABLE - 2

A N O R E C T A L A N O M A L I E S

1980 to June 1 9 9 1

TOTAL: 3 3 2

M A L E	F E M A L E
229	103

(a) Vestibular Malformations	69
(b) Perineal Canal	7
(c) Perineal groove	1
(d) Cloaca	16
(e) Recto Vaginal fistula	6
(f) Congenital Short Colon	1
(g) Anal Stenosis	3

EMBRYOPATHY OF RECTO-VESTIBULAR FISTULA:

Internal and external cloaca are separated by Cloacal membrane. A TOURNEAUX FOLD form a saddle shaped cleavage of the cloaca as far distally as the 'MULLER HILLOCK'. Rathke Folds fail to appear. The Mullerian ducts descend upon the partially devided cloaca. The Mullerian ducts then migrate down the posterior wall of the undivided cloaca carrying the attenuated rectal fistula and orifice with them into the individual external cloaca. (STEPHENS F.D.,SMITH E.D. "Anorectal malformations in Children" Chicago year book Medical Publishers - 1971). The anterior perineal anus results from inadequate sagittal length of the perineal mound in the uro-anal septum. The perineum between the anus and the fourchette is foreshortened. The position of the vulvar anus results from almost complete lack of development of the same septum, the perineum is absent, the fourchette rudimentary and the borders of the vestibule elongate posteriorly to accommodate anus.

The formation of anocutaneous fistula occurs when the medial migration of the genital folds occurs in a great extent and the anus is covered over by the folds and its lumen is projeced anteriorly on to the perineum or fourchette to form the ano-cutaneous fistulae.

DIAGNOSIS AND INVESTIGATIONS IN VESTIBULAR MALFORMATIONS:

All female babies born with absent anal canal should be examined under proper illumination. The perineum should be carefully examined using a probe or a catheter to establish communication of the "Hind Gut" to the vulva, vestibule, vagina or perineum. If the probe goes vertically upwards from the external opening into the rectum a recto vestibular fistula is diagnosed. When the probe goes posteriorly into the anal canal a communicati between the anus and the opening at the vestibule or vulva is established. A fistulogram will demonstrate the level of the anorectal canal and the length of the fistula. CAT scan of the Pelvis helps in estimating the development of the levator and the sphincter muscles . To diagnose associated anomalies, specific investigations like urine examination, serum creatinine, ultrasound of the abdomen, IVP, micturating cystourethrogram, cys Chest and Spine X-rays and other investigations as indicated on clinical examination are necessary.

MATERIAL AND METHOD:

A N O V E S T I B U L A R F I S T U L A

	Age at Presentation	Age at Operation
Neonates	6	-
1 month - 1 Yr	18	17
1 Year - 2 Year	27	26
2 - 5 Years	4	4
> 5 years	14	13
Total:	69	60

The babies with anovestibular fistulae are usually diagnosed early and brought for management during neonatal period or during infancy. Many are brought after the weaning period due to constipation. A few may present with partial obstruction when the fistulous opening is narrow. An abnormal opening in the genitalia or perineum is usually noticed by the parents Our policy in management of vestibular malformations is established during the past three decades. We prefer to dilate the opening at the vestibule or vulva in all Low Anomalies.

This is followed by anal transposition when the baby is about 4-5 months of age. A preliminary colostomy may be required in few children who have very narrow fistulous opening before embarking on anal transposition. Colostomy is also indicated when the babies present with obstruction due to faecaloma, redo operation, malnutrition and in children above 2 years of age.

Anal transposition can be performed by anterior approach in the lithotomy position or through a sacroperineal approach in the Jack-knife position.

Initial cut back operation to relieve constipation may be required in a few when the opening is narrow and not amenable to dilatation. Cut back anoplasty as a definitive procedure for anovestibular fistula is not recommended as the post operativ appearance of the perineum is aesthetically not acceptable to the patient as well as the parents. 'Z' plasty and rotation flap-perineo-plasty have been used in some centres for correction of anovestibular fistulæ, claiming satisfactory results.

For the intermediate variety of rectovestibular fistulae no cut back or dilatation is advocated as the fistulous tract is long. A prompt colostomy could be established in the early

neonatal period. A definitive operation can be performed when the baby weighs about 7-10 kilograms. Sacroperineal or a posterior sagittal approach for correction of this recto-vestibular fistula gives fair to good results. The colostomy can be closed in 2-3 months after definitive surgery.

RESULTS OF ANAL TRANSPOSITIONS:

In assessing the results of anal transposition four important criteria are taken in to consideration. 1. Anorectal continence 2. mucosal prolapse, 3. anovulvar distance and 4. constipation. Modified Wingspread method of assessment is simple, and can be used in any situation where sophisticated manometric studies are not available.

CRITERIA FOR ASSESSMENT OF CONTINENCE

WINGSPREAD METHOD (MODIFIED)

Clean Smearing	..	Good
Intermittent Soiling	..	Fair
Constant Soiling	..	Poor

In this method of assessment, children who remain clean as well as those who have smearing of faeces are grouped under the category "Good". If they have intermittent soilin the results are considered "Fair". They are assessed as poo when there is constant soiling.

In our assessment of 42 patients after anal transposition in 85.7% had good results and 14.3% had fair results. There were no patient with poor results. Mucosal prolapse tends to be slightly increased in the sacroperineal approach than in the anterior approach. This could be due to excessive mobilisation of the rectum through the sacroperineal route The anovulvar distance was adequate in all who had primary healing after anal transposition. Minor degrees of constipa are seen in few patients. They are managed satisfactorily with laxatives whenever required.

PERINEAL CANAL AND PERINEAL GROOVE:

Perineal canal is one of the rare vestibular malformations. The anal canal is at the normal site or it may be slightly anteriorly placed. The anorectal sphincters are well develo but there is a communication between the anal canal and vest anteriorly. Many consider this as a congenital abnormality they present during the neonatal period or during early infa

The perineum is a composite structure formed from the primitive perineal mound and genital folds. When the mound contribution is minimum and the genital folds assumed a major role and migrate medially a canal is formed between the anterior wall of the anus and vestibule. If the genital folds fail to migrate medially and the mound contribution is made a perineal groove is formed. This is the explanation given for congenital development of perineal groove and perineal canal. Those who consider this as acquired lesion states that the perineal canal develops after rupture of an abscess between the anal canal and vestibule establishing a fistulous communication.

Children with perineal canal present with excoriation of the skin around the vestibule due to constant contamination with faeces and infection. They are managed conservatively with antibiotics and local cleaning in the initial period.

Often if the infection is severe, a temporary colostomy may be required, to control the infection. The fistulous tract can be excised after control of infection and colostomy closed subsequentl If the infection is controlled without colostomy, the fistulous tract may be excised primarily. The tract should never be laid open as it will require repair of the anal sphincter and perineum.

PERINEAL GROOVE: can be corrected by excising the mucosal lining of the groove and closure of the defect with surrounding skin.

CONCLUSION:

Anal transposition is advocated for good aesthetic and function results. The patient as well as the parent are satisfied with the results of anal transposition. Colostomy may be required as an initial procedure or along with anal transposition in older children or while re-doing the operation. Perineal canal requires complete excision rather than fistulotomy.

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A PERSISTENT CLOACA

Dr. S. VASANTHAKUMAR

A persistent cloaca is defined as the conjunction of the rectum, vagina and urethra with a single common channel (cloaca) and represents 10% of the entire group of anorectal malformations. The main goal in the treatment of cloaca is to achieve bowel and urinary control by accurate separation of rectum from the vagina and vagina from the urinary tract.

A proper evaluation of the urinary tract in a case of persistent cloaca is to be stressed as severe obstructive uropathy is frequently associated with morbidity and mortality.

So a newborn with persistent cloaca may require emergency decompression of the urinary tract in the form of cystostomy, a ureterostomy or a pyelostomy or vaginostomy even prior to establishment of a colostomy. One study has shown that 68% of these patients had important associated urological defects such as severe hydronephrosis, vesicourethral reflux or neuropathic bladder. Hydrocolpos and different degrees of vaginal and uterine septation have been associated with persistent cloaca.

The repair of the entire malformation may be undertaken after waiting for 6 months, during which period the entire urinary tract may be evaluated.

Diverse treatments have been proposed in the past for the management of this defect. The traditional approach was to repair the rectal component of the malformation, having the genitourinary sinus part for a secondary operation at a later time. The new approach called posterior sagittal anorecto-vagino-urethroplasty (PSARVUP) allows direct exposure to the complex visceral arrangements as well as the voluntary muscle relevant to urinary and fecal continence. This approach is the most ideal approach to treat this wide spectrum of defects with different degrees of complexity.

The details of the surgical technique have been described by Alberto Pena. I like to stress that use of electrical stimulator helps in accurate checking of the muscle on both sides of midline and magnification helps the surgeon to separate the rectum and vagina which have a common wall. The vagina and urinary tract also share a common wall and these two structures are less elastic and also because the common wall is not flat, since the vagina wraps in a semicircle (180°C). the post urethral wall and bladder neck.

The common channel is then reconstructed in two layers around a foleys catheter to create a neourethra. The vagina is located posterior to the urethra and is sutured to the perineum. The rectum is located in front of the levator muscle immediately behind the vagina in the upper part. The lower part of the rectum curves posteriorly and the new anus is located at the centre. The posterior edge of the muscle complex is sutured together anchoring the posterior wall of the rectum with the same suture to avoid post operative rectal prolapse.

Additional technical manoeuvres may be needed either because the rectum is found too high and must be mobilised from above or else because the vagina does not reach the perineum and it may require a piece of intestine. Common Channel (cloaca) more than 3 cm. in length requires some of the technical alternatives to replace the vagina. These include perineal skin or labial flaps, vaginal replacement with small or large bowel in cases of short vagina with large dome (as seen in cases of hydrocolpos). One can rotate down a tubularised flap of the upper vagina to built the lower vagina. Foleys catheter is left in place for 2 weeks or do not hesitate to create suprapubic cystostomy when considered necessary to protect the anastomosis.

We have had 3 cases of persistent cloaca reconstructed during the last 4 years by the technique of PSARUVP. In one case the rectum was ending as a long fistula requiring the abdominal step mobilisation of the bowel. All the 3 cases had short common channel and reconstruction of the vagina was carried out using either perineal or labial skin flaps. All the three underwent colostomy on the 2nd day of life and did not present with obstructive uropathy or hydrocolpos. During the waiting period the urological work up was done which included ultrasound, genitogram and genitoscopy. There was no abnormality in the sacral vertebrae. The definitive surgery was done at 1½ years. Colostomy was closed 3 to 4 months after the definitive surgery.

The number is small and our experience is limited to these 3 cases where they did not have abnormalities of the urinary tract or skeletal system. The reconstruction of vagina was feasible due to the short wide common channel. We have not had an occasion to use the large or small bowel to reconstruct the vagina. All the 3 patients have fairly good continence, soiling only occasionally.

When compiling results one must be specific as to the type of cloaca being dealt with. The main prognostic factors are quality of the sacrum, quality of the muscles, length of the common channel, size of the vagina and the height of the rectum.

There is a significant difference in clinical results between patients with normal Vs abnormal sacrum. Patients with abnormal sacrum were most frequently associated with poor muscles. Long common channel is also associated with poor sacrum.

PSARVUP requires a great degree of patience, meticulous technique and should be performed by a center that has had the opportunity to do enough of the procedure so that sufficient expertise has been developed.

COLON PREPARATION

Dr. V. KALIDASAN

"For want of a shoe-nail the battle was lost"

Preparation of the colon is essential before :

i) Anorectal reconstruction ;
ii) Colostomy closure.

Before anorectal reconstruction there are two parts of the colon to be prepared : a) the proximal colon and b) the distal blind pouch. Preparing the proximal colon is essential because of two reasons :

i) Occasionally it may becc me necessary to close the colostomy and undo it to use it as part of the pulled- through bowel;

ii) A well prepared proximal colon prevents any form of contamination of operative areas during surgery and also prevents spill over into the distal pulled-through bowel in the immediate post-operative when it is all important to keep the suture lines clean.

Preparation of the proximal colon is similar to that done before colostomy closure and is discussed below.

Cleaning of the blind pouch should begin immediately after colostomy is performed in the neonatal period. Some surgeons prefer to start this even on the operating table at the time of colostomy. Keeping the blind pouch clean is important because it prevents meconium in the pouch from becoming inspissated and also reduces the incidence and severity of the urinary tract infections that occur due to contamination through the recto-urethral fistula. The blind pouch is thoroughly washed out in the neonatal period after colostomy is performed preferably with warm saline. When the child is discharged after colostomy the blind pouch should be empty of meconium. Needless to say the cleaning of the blind pouch is easier if sigmoid colostomy has been done.

However, when loop colostomies have been done a certain amount of spill from the stoma to the distal bowel is unavoidable. This has to be got rid off by thorough wash-outs at the same time as the proximal bowel is being prepared. Preparation of the proximal bowel involves the following :

i) Dietary modifications
ii) Mechanical cleansing of the bowel
iii) Antibiotic prophylaxis.

Dietary modifications are not warranted when the child is an infant on liquid diet. Such modifications become necessary when the older child on a mixed diet is coming up for surgery. The child is put on soft solids of a low-fibre, low-residue type 48 hours before surgery for 24 hours. During the last 24 hours he is kept on clear fluids only orally. It is important to take into account his fluid, calorie and electrolyte requirements during this time, especially because he is also likely to get colon wash-outs etc, during this period and dehydration or electrolyte disturbances should be prevented. During the two days prior to surgery he is given a oral laxative at bedtime. Total bowel preparation 6 to 8 hours before surgery (by producing diarhoea) can be achieved by making the child ingest 300 to 500 ml of a 10% solution of mannitol or an isotonic saline solution (2 to 4 litres). However, this is not a popular method in children because of the discomfort involved; it is also dangerous because of the possibility of producing significant dehydration and serious electrolyte disturbances. Colonic washouts are the easiest and safest. Either saline or tap water may be used. The secret of success in having a clean colon is a committed nurse or resident. The washouts have to be given patiently and each time the wash has to be continued till the return is clear. Large quantitities have to be used to successfully wash the proximal colon. Colon washouts are given 36 hours, 24 hours, 12 hours before surgery and again on the morning of surgery.

Antibiotics are started orally 24 hours before surgery and parenteral antibiotics are given along with the pre-medication before surgery. The regimen we follow for colon preparation before colonic surgery is given below :

Admit child atleast 48 hours before surgery.

Soft, low residue, low fibre diet started on admission.
(48 hours prior to surgery)

Liquid diet started 24 hours prior to surgery.

Dulcolax orally (or cremaffin liquid) given on night of admission and on night before surgery.

Colon washouts given on night of admission (36 hours before surgery), next morning, night before surgery and morning of surgery.

Oral erythromycin and metronidazole started 24 hours before surgery.

Parenteral gentamicin, ampicillin and metronidazole given one hour prior to surgery.

The importance of Colon preparation lies in the fact that a badly prepared colon can lead to poor post-operative results. Faecal leaks, faecal fistulae increase the morbidity and occasionally can even lead to death. It is a disaster to close a child at the time of colostomy closure when he has weathered the trauma of a neonatal colostomy and a later pull-through. Therefore, if poor bowel preparation is the cause of failure, it is indeed a case of losing the battle for want of a shoe-nail.

Mechanism of Anorectal Continence in the Normal and Measures to Preserve it in Anorectal Malformations

DR. T. DORAIRAJAN

ANORECTAL CONTINENCE

DEFINITION "Anorectal Continence is the ability to retain voluntarily or involuntarily the intestinal contents until an evacuation is desired"

HOLSCHNEIDER

"The attainment of perfect post-operative continence is the goal of any surgery for imperforate anus - TAILOR et al 1973."

Anorectal Continence is a vital and complex function in the human being making possible integration of a person into society. Faecal incontinence is a grave handicap to social acceptance. This vital function has been studied in health and disease. The interested reader has to consult the contributions of SCHARLI and KIESSEWETTER (1969), VARMA and STEPHENS 1972, VARMA 1972, STELZNER 1976 and HOLSCHNEIDER 1976, 1988, SCHARLI 1989.

The rectum and anus with their envelope of sphincters erect a series of barriers to evacuation of intestinal contents. These barriers have to be surmounted and a conscious effort of straining has to be made before the gut can be emptied of its contents, when the urge to defaecate occurs. The sum total of these barriers to evacuation constitute the Mechanism of Anorectal Continence.

The ability to retain contents is ensured by the following :

I. The Rectum)
II. Motor) factors
III. Sensory)
IV. Reflex)

I. ROLE OF RECTUM : The normal rectum has an important role in anorectal continence by its specialised properties. The rectum impedes unguarded descent of its contents by the following mechanisms :

1. Rectal pressure gradient
2. Rectal adaptation and compliance
3. Presence of Houston's folds
4. The anorectal angle

The Rectal Pressure Gradient : In the resting stage, the rectum keeps itself empty by increased muscular activity that creates a high pressure zone whose pressure is higher than the pressure in the sigmoid colon.

The slow wave electrical frequency of the rectum is between 18-24 cycles/min. while that of upper rectosigmoid region is 4-8 cycles/min. Because of the difference, small quantities of contents are pushed back in the oral direction in an antiperistaltic manner. Thus small frequent leaks are prevented.

Rectal adaptation reaction and compliance : This is an important physiological property of the rectum that ensures continence. When the volume of rectal contents is increased, there is a sharp increase in intra-rectal pressure followed by slow decrease. This property is termed adaptation reaction. This is the first defence against faecal leak and ensures that intra-rectal pressure does not exceed sphincter resistance.

Compliance is the property that denotes elasticity of the rectum.

Rigid rectum is less elastic and less compliant and hence will cause incontinence. Small quantities of faeces will raise the pressure sharply and overcome sphincter resistance, resulting in leakage. Rectum with high compliance will delay passage of contents and cause constipation.

The junction of the rectum and anus is acutely angulated forward and laterally compressed by the puborectalis fibres. Houston's folds which are three fixed mucosal invaginations impede unchecked descent of formed stools by gravity.

II MOTOR FACTORS :

The Puborectalis part of the Levator ani
The deep and superficial external sphincters
The internal sphincter

The Puborectalis muscle plays a dominant role in anorectal continence. This 'U' shaped sling like muscle draws the anorectal junction forward and by its tone, angulates the anorectal junction and occludes the lumen.

The External anal sphincters : The deep and superficial parts of the external sphincters which are striated muscles, act as a functional unit in ensuring continence by keeping the anal canal closed unless defaecation is desired.

The internal sphincter which is smooth muscle, by its resting tone, keeps the anal canal closed. Its contribution is estimated to be 10-12 mm of Hg. 20 mm of Hg pressure are contributed by the striated muscle sphincters (Puborectalis and external sphincters).

The combined sphincter muscles (smooth as well as striated) on an average exert 30 mm of Hg. pressure.

III. SENSORY FACTORS : (VARMA, STELZNER) Sensory perception of changes in the rectum and anal canal are vital to anorectal continence. Changes in intra-rectal pressure, the nature of contents (flatus, fluid and faeces) are accurately perceived and bring forth automatic unconscious and conscious responses. Three zones of sensory perception are recognised.

a. **The rectal area :** Acute distension of the rectum gives rise to a sense of fullness which is vague in localisation but unmistakable in warning about the threat to continence.

b. **The sphincter area** : This is equally vital in sensory perception.

Stretching of the striated fibres of the puborectalis as well as the anal sphincter give acute desire to defaecate. This sensibility is not lost after resection of rectum and substituting the sigmoid colon for pull through. SCHARLI has called this area "the trigger zone of continence".

c. **The anal area** : The epithelium of the anal canal perceives sensations as to the nature of contents.

IV. REFLEX FACTORS

Effects on sphincters of raising intra-rectal pressure : Raise in intra-rectal pressure has complex effects on the internal and external sphincters.

The internal sphincter relaxes when the intra-rectal pressure is raised. This is termed RECTO-ANAL reflex.

In the case of external sphincter, the effect is more complex. There is initially an involuntary reflex contraction of the voluntary sphincter, followed by a conscious contraction of the voluntary sphincter. VARMA and STEPHENS have termed this PHASE I and PHASE II reaction. SCHARLI terms these reflex responses, the Continence reaction.

When intestinal contents descend into the rectum, the sensation of desire to defaecate is perceived by the cortex due to the stretch of receptors in the para-rectal muscle complex (Puborectalis and levator ani). The intra-rectal pressure is sharply elevated. The internal sphincter relaxes. The slight descent of contents into the lowest part of rectum and the uppermost part of anal canal (the TRIGGER ZONE of continence of SCHARLI) causes the external sphincter promptly to close with greater force till it is overcome by the conscious act of straining to defaecate or consciously the force of contraction of external sphincter may be reduced to enable evacuation.

CONTINENCE AFTER SURGERY FOR ANORECTAL MALFORMATIONS The degree of continence in anorectal malformations depends on the type of anomaly.

Low malformations have near normal continence since the various factors responsible for continence are not affected.

In high malformations the following motor and sensory components of continence are affected. The internal sphincter as an effective structure is absent and rudiments are present near the recto-urethral fistula and its quantitative contribution to continence is negligible or absent.

VARMA and STEPHENS after sacroperineal reconstruction for high anomalies could not demonstrate Phase I reflex sphincter contraction.

Factors in surgical technique which will influence continence after Surgical correction: Extensive dissection and mobilisation of rectum will sever the afferent neural pathways responsible for continence. Dissection and mobilisation should be kept to the minimum.

The plane of dissection : Enveloping the distal rectum in high anomalies is the visceral layer of pelvic fascia. All dissections should be made within this fascial envelope.

The rectum and perirectal tissues should be kept supple and scar free by maintaining good blood supply and avoiding infection. Rigid rectum or perirectal fibrosis will alter rectal compliance and rectal adaptation.

Hence the need for minimal and gentle dissection and mobilisation.

Accurate tunnelling of the rectum through the puborectal sling : This is the important step that will ensure continence. The puborectalis sling closely surrounds the urethra and lies below the recto-urethral fistula. It is funnel shaped, wide above and narrow below. The space between the sling and the urethra has to be dilated gradually with Hegars dilators prior to pull through.

Bulky rectum requires attention by tapering, if necessary. After pull through, the mucosa of the pulled bowel should be pink. If it is dusky, it means there is venous compression. Dilatation gently through the lumen at this stage helps. If the mucosa is totally pale and white, it means vascular compromise. This can be due to tight stretching of the pedicle or tight squeeze by the sling. In either case, the bowel should be pulled out and after restoration of blood supply, the error rectified before pulling through again.

These technical details spell the difference between success and disaster. The lowest portion of bowel to be pulled down is not usually the site of fistula. The fistula usually opens slightly higher ($\frac{1}{2}$ - 1 cm).

The Anal sphincters in high malformations In the classical sacro-perineal proctoplasty of STEPHENS, the external sphincter is not mentioned as of any use.

However, the recent work of DEVRIES and PENA and the regular use of electrical stimulation to identify contractile muscles and the experience of other surgeons including the author indicate that there is extension of the sphincter mechanism caudal to the puborectalis fibres. Their quantity and disposition is variable. But all efforts must be made to identify and place the bowel within their grasp. Perianal soiling and mucosal prolapse that distress the patient can be avoided by inverting the perianal skin by the methods of NIXON and CYWES.

Supple and scar free skin-mucosa junction:

This is essential for normal anorectal function. Skin-mucosa junction strictures produce painful and incomplete evacuations which alter continence. They are prevented by designing suitable flaps. A fair proportion of children require dilatation. But the surgeon should know when dilatation is insufficient and secondary procedures are necessary to release strictures. Scar excision, "Z" plasty, rotation flaps and other procedures can permanently relieve refractory strictures in this region.

After reconstruction degree of continence improves with passage of time:

The degree of continence achieved soon after reconstruction of high anorectal anomalies, is satisfactory in only 30% or less of children. However, this percentage slowly increases with passage of time. The time taken to achieve acceptable levels of continence is variable in a given child and may in some cases take as long as ten or more years. This is attributed to several factors :

(a) The child's awareness of the need to be continent.

(b) Lengthening of the sphincter zone by growth. A minimum of 3 cms of sphincter is necessary for ensuring continence.

(c) "Maturation of Rectum"-a concept introduced by HOLSCHNEIDER to indicate the development of reflex sensibilities of the pulled bowel, whether rectum or sigmoid colon, with passage of time.

The writings of NIXON refer to relationship between age at reconstruction and speed of development and efficacy of continence. Infants undergoing neonatal reconstruction or within three months of birth fare much better than babies reconstructed at or after one year of age.

SUMMARY ;

In the human being the evacuation of faeces is barred by a series of mechanisms, unless consciously surmounted when the desire to defaecate is felt.

Craniocaudally these are :

1. Rectal pressure gradient
2. Rectal adaptation and compliance
3. Acute anorectal angle
4. Combined tone of internal sphincter, Puborectalis and external sphincter.
5. The reflex properties of all these muscles.

Combined together the sphincter mechanism exerts a pressure of 30 cms of Hg. Only when this is overcome, defaecation is possible.

In high anorectal malformations, the internal sphincter, as an effective organ, is absent. The surgeon by his technique has to utilise available puborectalis fibres and the external sphincter mechanism. The anorectal angle has to be created. The reflex mechanisms are deficient. They will be further curtailed by extensive dissection, and handling.

Tapering will alter compliance and rectal adaptation. It has to be avoided or kept to the barest minimum.

A skin lined proctodeum by inversion proctoplasty, and a supple scar free skin-mucosa junction have to be ensured.

Finally the reconstructed lower end of gut with its sheath of available sphincters requires considerable time to reach functional maturity. Early reconstruction before three months has superior functional results compared with later repair.

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Anorectal Anomalies - A Retrospective Study

Prof. P. N. NAGENDRAN
Dr. HAYAVADANA RAO
Dr. VIVEKANANDAN

This is a retrospective analysis of 75 cases of Anorectal Malformations which were treated at the Department of Paediatric Surgery, Institute of Child Health & Hospital for Children over a period of one year (1989-90). These cases were treated by five Unit Chiefs, 9 Assistant Professors & 6 M.Ch. Post graduates.

TABLE I

Types of Malformations

High	-	55
Intermediate	-	6
Low	-	8
Cloacal Anomaly	-	6
Total		75 cases.

TABLE II

Sex Incidence

Female	-	16
Male	-	59

TABLE III

DETAILS OF TYPES OF ANOMALIES

High		
	ANORECTAL AGENESIS	
	Without fistula	24
	With Fistula	25
	Rectal Atresia	6
INTERMEDIATE		
	Anal Agenesis	2
	Anovestibular fistula	4
Low		
	Anocutaneous fistula	8

Following associated Anomalies were noted :

TABLE IV

Systemic (Down's syndrome)	2
CVS	3
Gastro intestinal	1
Genito Urinary	7

TABLE V

Types of Urological Anomalies

Hypospadias	2
Hydronephrosis	1
Double Ureters	1
Non Refluxing Megaureter	1
Vesico Ureteric Reflux	2
Total	7

63 Children had colostomy

Transverse Colostomy	29
Sigmoid Colostomy	34

The following types of Operations were performed for high and intermediate malformations.

PSARP	42
SACRO ABDOMINO PERINEAL	6

The following complications were noted :

TABLE VI

COMPLICATIONS

Recurrent Recto Urethral Fistula -	3
Bladder Injury	3
Urethral Injury	1
Stricture Urethra	2
Anal Stenosis	2
Rectal stricture	2

Mortality was 3 out of 75 children

Review of Current Literature on Anorectal Anomalies

Dr. V. R. RAVI KUMAR

Though literature on anorectal anomalies (ARA) is vast several questions regarding the management of high and low anomalies and assesment of continence and the long term results are continuously being evaluated by different centres dealing with ARA. In the ensuing article the long term results of Posterior Sagital anorectoplasty (PSARP) and the time tested Sacroperineal (SP) and Sacroabdominopeineal (SAP) procedures are analysed, as reported from different centres. The value of CT and MRI scans in diagnosis of ARA and assesment of placement of rectum in relation to the striated muscle complex (SMC.) are highlighted from two different centres. Results of PSARP done in neonatal period as one stage procedure are discussed. Different methods of assesment of continence by different clinical systems like Kelly, Templeton, Kiesewetter and Wingspread group are discussed.

Pena and de Vries introduced Posterior sagital anorectoplasty in 1982 as a new approach for treatment of ARA. The advantage claimed was PSARP allows direct exposure of anatony of these defects, allows separation of Rectum from genitourinary tract without damaging important structures. This approach as per Alberto Pena has important implications for therapy, evaluation of results, classification and terminology. Alberto Pena discussing the contreversial issues and criticisms against PSARP answers the basic issues involved against PSARP.[1]

The main criticisms are 1. Potential irreversible damage to puborectalis sling (PRS) by cutting it and also that sutures do not hold the muscle well. The reluctance on the part of surgeons to cut the puborectalis is based on the conventional anatomic knowledge that the PRS goes around the urethra and hence the reluctance to cut it. Pena explains that the muscles around the rectum run in cephalcaudal direction with very few muscle fibres are horizontal. So one is actually separating the parallel muscle fibres which fall back and hold the sutures well behind the pulled rectum.

The second criticism is that tapering may provoke severe non-functional rectal stenosis. The rectal stenosis is primarily due to devascularisation of the rectal stump more common in prostatic and vaginal fistula and not due to tapering. The strictures are more often at the bowel and skin level which responds to dilatation. Many surgeons were worried about the denrervation of the pulled intestine in PSARP while dissecting to gain length and many develop chronic constipation. Since conventional techniques also need mobilisation, there is no foolproof technique in which the most distal part of the rectum is pulled down with its innervation intact.

Can PSARP incision and exposure could be used to create a tunnel in front of the PRS without actually dividing the muscle? This Pena considers as a compromise by traditionalists which may result in lateral and anterior mislocation of rectum and damage to delicate muscle fibres. Durham Smith suggests that for all but very high lesions the de Vries-Pena exposure is the optimum starting point but with preservation of Puborectalis, Pubococcygeus and the deep portion of the external sphincter intact (NOT DIVIDED).[2]

Since there is a lot of overlap of what is considered a high, intermediate or low anomalies Pena suggested that for a assesment of long term results specific defects with anatomical characterstics should be compared. Since the lower limit of levator ani cannot accurately be defined the concept of infra levator malformation according to Pena is questionable and more than 90% of anorectal malformations are trans-levator, as majority of the bowel lies somewhere in the levator tunnel.

In a retrospective evaluation of functional results of 332 patients by Pena (161 patients were excluded) operated between 1980-1987, patients were grouped according to the potential for continence.[3] Voluntary bowel movements (VBM) were considered as one of the best indicaters of faecal continence and was found in 77% of patients with normal sacrum and rectourethral fistula. VBM was only 30% in children with rectovesical fistula. All patients with low malformations and normal sacrum had voluntary bowel movements. Surprisingly the incidence of soiling is 20% with Vestibular fistula and 6.1 % with rectourethral fistula. The incidence of soiling is highest with Cloacal anomalies (75%). With Sacral agenesis of 3 sacral vertebrae the voluntary bowel movement was present only in 20%. Reoperation by PSARP by Pena for fecal incontinence who were cperated elsewhere showed marked improvement in 45% of cases, some improvement in 37% and no improvemant in 18% of cases. Reoperation in abnormal sacrum achieved marked improvement in only 20% of cases. The patients were considered as having voluntary bowel movements when he or she was able to tell the mother that he or she wanted to defaecate and was able to have a successful bowel movement without the help of enema or suppository.

The objective methods of measuring the bowel function such as defaecography, rectal monometry, electromycgraphy. C.T. and MRI scans are well known and Pena is of opinion that these studies do not show good correlation with clinical picture and quality of life. He is of opinion that excellent manometric results were sometimes asscciated with poor quality of life due to constant soiling. K.K. Varma[4] found a direct correlation between manometric assesment and clinical assesment. Further manometry allowed a prognosis to be established and reoperation determined. R. Rintala[5] found that presence of rectoanal inhibitory reflex correlated positively to faecal continence. Continence in patients with positive rectoanal inhibitory reflex was significantly better than in whom the reflex was missing.

It is observed that patients with high malformations who require laparotomy for complete repair have much poorer prognosis as regards anal continence even though the sacrum is normal. It is suggested that poor prognosis is due to poorer musculature. Children who had Sacroabdominoperineal (SAP) surgery showed lesser continence levels compared to purely Sacroperineal procedures. The manometric studies by K.K. Varma showed that good maximal squeeze pressure in children who had SAP and the reason for lesser continence was thought to be the lack of preservation of rectal innervation.

In a review of 50 patients who underwent PSARP for high ARA between 1983 to 1990 Richard et al from Netherlands reported that subjectively the majority of patients were more or less incontinent with soiling of clothes atleast once a day. On the basis of objective criteria virtually all patients appeared to be incontinent from that study they concluded that although PSARP provides good esthetic result, patients will never acquire normal continence. (The assesment has been made by anamnesis (subjective), elector stimulation, defecography, and anal monometry.

A study of the long term continence in patients with high intermediate anorectal anomalies treated by Stephens sacral approach, was published with a follow up of 5 to 32 years at the Royal children's hospital, Melbourne.[7] The continent levels were analysed by Kelly, Templeton, Kiesewetter and Wingspread methods. The overall results summarised show that the proportions of patients in continence category were similar in Kelly, Kiesewetter, and Wingspread methods of assessment of anal continence. The review demonstrated that 65 to 90% of patients following SP and SAP rectoplasties attained socially acceptable continence. As in other studies the continence rate of SP is better than SAP. The continence rate in females is higher than male children, the reason being male children have higher incidence of High anomalies. Female children tend to have intermediate anomalies with better pelvic musculature.

Do the children with ARA continue improving in faecal continence as they grow older? It is argued that improvement with age is due to the ability of the patient to adjust their diet, drugs, enemas etc, and achieve a social adaptation. Varma found that those who had a MSP (Maximal Squeeze Pressure) of over 40mm of Hg improved in continence over a period of years. Those with poor MSP fared poorly. It was observed that if the sphincter is left undamaged, improvement can be expected after upto ten years. The improvement is seen between 5 and 10 years.

In a follow up of 37 patients over a period of 5 to 32 years from Royal Children's Hospital, Melbourne 9 out of 37 patients reported improvement in continence between 7 to 15 years. It was speculated that the late onset of faecal control is likely to be the result of multiple factors including increased muscle mass control of pelvic musculature, regulation of diet, education & social motivation.

Nai Theow Ong and Spencer W Beasly [8] of department of surgery Royal Children's Hospital, Melbourne have assessed the continence after repair of high ARA by different clinical methods by Kelly, Templeton, Kisewetter and Wingspread Methods. It was concluded that Kelly scoring system has the advantage of being simple and universally accepted. It correlates well with radiological, manometric, electomyographic measurement of continence though it necessitates a rectal examination. (Table 1 and 2)

What are the long term functional results after perineal surgery for low ARA?

70 patients with low anomalies treated by perineal operations were followed up from 5 to 38 years with a mean of 16 years. Over 90% achieved socially acceptable control.[9] The incidence of accidental defaecation has a lower incidence after 10 years. The incidence of smearing is 44% and soiling 24%. The reasons attributed to such figures are damage to sphincter during cutback, inadequate cut back and congenital deficiency of the muscle mass. The incidence of soiling is only 14% in children more than 10 years old.

Repair of ARA in neonates by PSARP : H. K. Goon[10] from Malaysia had operated on 33 neonates with ARA by PSARP with no mortality with few minimal complications. Tapering of the bowel was not done as in the conventional PSARP. The result as per the author is good after a follow up of 1 to 3 years. However the follow up period is too short to assess the functional result. Out of the 33 neonates only 3 had high lesions, rest were intemediatre and majority were in female children.

Is there an internal sphincter in ARA?

Rosto Rintal[5] studied two groups of children operated for ARA for internal sphincter function. In the first group rectal blind pouch and the fistula were utilised in the reconstruction of anal canal and in the second group the sigmoid and proximal colon were pulled through to the anus by SAP. The feacal continence in the first group was superior to the second group. The findings show that in anorectal malformations there is internal sphincter function in the distal rectal pouch and that it can and should be preserved when anal canal is reconstructed. In a study on rats, transplacentaly induced anorectal malformations showed smooth muscle hypertrophy at the rectal end which seemed to be considered as internal sphincter.[11]

What is the role of CT and MRI in ARA?

Localisation of blind rectal pouch is conventionaly done by the radiograph taken with the infant in upside position. The CT scan has the potential to identify the level of blind rectal pouch soon after birth without waiting for the air to reach the blind end.[12] MRI[13] not only delineates muscle complex accurately but identifies spinal and spinal cord abnormalities. MRI is useful especially in secondary cases to determine the initial failure. The lack of adequate pelvic musculature or poor placement of distal colon in relation to SMC or the presence of Mesenteric fat can accurately be delineated. Unsuspected lesions such as tethered cord, sacral abnormalities and urinary tract anomalies can be recognised. In the new born it gives a very clear imaoge of rectal pouch.

Percutaneous transperineal pouch localisation for low anal anomalies is described by Richard et al.[14]

Using a muscle stimulator the centre of the anal dimple is marked A 22 gauge needle was introduced into the rectal pouch. The position of the needle was confirmed and 10 F intravenous cannula was passed into the rectum. The metal needle was removed and 6 F FOGARTY catheter was introduced through the sheath of 10 F cannula. Through a cruciate incision with Fogarty catheter at the centre and with traction to the Fogarty the blind rectal pouch can be identified and it facilitates easy dissection with minimal damage to muscle fibres.

Summarising, the results of surgery by Pena and de Vries could not be reproduced by other surgeons doing the same procedure. Review of results by Stephens method and long term follow up done in various centres have shown good results for continence which seems to improve with age. The results for SP is better than SAP. The necessity to preserve the distal blind end of the rectum or the divided fistula for reconstruction is emphasised to preserve internal sphincter function. PSARP done in neonatal period has given good results with no mortaility thcugh the period of follow up is short. The C.T. and MRI scans are helpful in identification of blind pouch in the neonatal period soon after birth. They are useful for assessment of muscle mass and location of the placement of rectum in children in whom the continence is not upto the mark A. New technique for localisation of blind pouch in low anal anomalies using a Fogarty's catheter which facilitates further dissection had been described.

Table 1. Summary of clinical scoring methods

1. Kelly method

Frequency of staining of smearing	None	2
	Occasional	1
	Constant	0
Occurrence of accidental defaecation or soiling	None	2
	Occasional	1
	Constant	0
Strength of puborectalis muscle action on digital examination	Strong squeeze	2
	Weak squeeze	1
	No squeeze	0

2. Wingspread method (modified)

Clean Smearing	Good
Intermittent soiling	Fair
Constant soiling	Poor

Table 2. Comparison of the two clinical scoring methods

Kelly score

1. Easy to remember and to use
2. Requires physical (digital) examination
3. Quantitive, allows progress charting
4. Based on promise that "good" score =normal
5. Correlates with some objective measurement techniques (defaecography, manometry)
6. Widely used and accepted method of assessment
7. Designed for use in any anorectal condition. e.g. Hirschsprung's disease
8. It is difficult for a patient with repaired anorectal anomaly to achieve a "good" continence score

Wingspread scheme

1. Physical examination not essential, but may provide information on faecal accumulation and rectal stenosis or prolapse
2. Descriptive, much more detailed than others
3. Importance of accumulation of stools highlighted
4. Diarrhoea not recognised
5. No definition of the frequency of "intermittent"
6. Grades the patient's dependence on therapy
7. Related complications noted separately
8. Cumbersome, not easily compared with other methods unless modified & simplified

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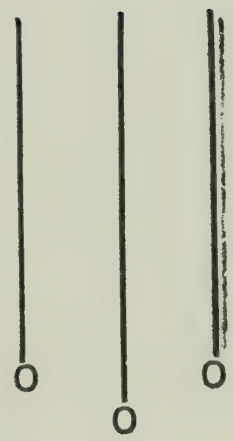

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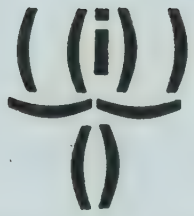

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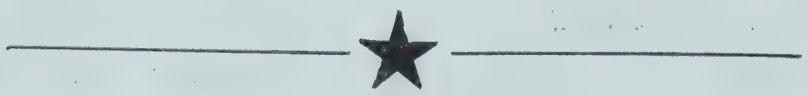

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